

DEFORMITIES OF THE SPINE

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Σπόνδυλοι δὲ οἱ ῥάχιν, ὅσοις μὲν ὑπὸ νοσημάτων ἔλκονται ἐς τὸ κυφόν, τὰ μὲν πλεῖστα ἀδύνατα λύεσθαι, ποτὶ καὶ ὅσα ἀνωτέρω τῶν φρενῶν τῆς προσφύσιος κυφοῦται...καὶ οἷσι μὲν κυφοῦται ῥάχιδες παῖσιν ἐοῦσι, πρὶν ἢ τὸ σῶμα τελειωθῆναι ἐς αὕξησιν· τούτοις μὲν οὐδὲ συναύξεσθαι ἐθέλει κατὰ τὴν ῥάχιν τὸ σῶμα, ἀλλὰ σκέλεα μὲν καὶ χεῖρες τελειοῦνται· ταῦτα δὲ ἐνδεέστερα γίνεται. Καὶ ὅσοις ἂν ἢ ἀνωτέρω τῶν φρενῶν τὸ κῦφος, τούτοις μὲν αἷ τε πλευραὶ οὐκ ἐθέλουσιν ἐς τὸ εὐρὺ αὕξεσθαι, ἀλλὰ ἐς τοῦμπροσθεν, τὸ δὲ στήθος ὅξυ γίνεται, ἀλλ' οὐ πλατύ, αὐτοὶ τε δύσπνοοι γίνονται καὶ κερχνώδεις· ἦσσαν γὰρ εὐχωρίην ἔχουσιν αἱ κοιλίαι αἱ τὸ πνεῦμα δεχόμεναι καὶ προπέμπουσαι.

When the spinal vertebrae are drawn into a hump by diseases, most cases are incurable, especially when the hump is formed above the attachment of the diaphragm. When hump-back occurs in children before the body has completed its growth, the legs and arms attain full size, but the body will not grow correspondingly as the spine; these parts are defective... Where the hump is above the diaphragm, the ribs do not enlarge in breadth, but forwards, and the chest becomes pointed instead of broad; the patients also get short of breath and hoarse, for the cavities which receive and send out the breath have smaller capacity.

Hippocrates, On Joints XL1 : 1-19

ABSTRACT

Deformities of the spine occur as a consequence of a number of different aetiologies each with a differing pathogenesis and natural history. The resulting deformity may be a scoliosis which is a lateral curvature of the spine in the frontal plane, a kyphosis which is an abnormal posterior curvature in the sagittal plane or a kyphoscoliosis which is a combination of both deformities. Significant progression of the curvature can occur with spinal growth and result in a very severe deformity with a major effect on general health, longevity and quality of life. If the deformity develops and progresses in early childhood, it can result in an impairment of lung growth and development possibly leading to *cor pulmonale* and death in early adult life. A kyphosis or kyphoscoliosis can cause spinal cord compression and if left untreated can result in paraplegia. Some patients develop back discomfort in later life, due to secondary degenerative arthritic changes, and others have a reduced self-image leading to psychological disturbances. Knowledge of the natural history is essential in anticipating problems and is the benchmark by which treatment is evaluated.

This collection of work is based on my studies of the aetiology, pathogenesis, natural history and management of patients with deformities of the spine seen over a period of 35 years while working as a spinal surgeon in Edinburgh. .

In 1975, I was appointed Senior Lecturer and Consultant Orthopaedic Spine Surgeon, working with Prof. J I P James in the Department of Orthopaedic Surgery, University of Edinburgh. It was here that I was able to establish a large database of patients, especially those with congenital deformities of the spine, who had previously received little or no treatment. This provided me with invaluable information and stimulated my lifelong interest in the natural history and allowed me to formulate an appropriate course of management for these conditions. In 1978, when Prof. James retired I took over his clinical practice and established the Edinburgh Spine Deformity Centre which later became, under my directorship, the

Scottish National Spine Deformity Centre treating all patients from the whole of Scotland.

At present there is virtually no severity of spinal deformity which cannot be significantly improved by surgery. However, it should be recognized that the necessity for surgical salvage procedures at a late stage, to correct severe deformities for conditions commencing as minor curves in infancy or adolescence, indicates a failure of management. This thesis emphasizes the need for early detection and prevention of severe deformity and depends on a thorough knowledge of the pathogenesis and natural history of the various conditions which can produce a scoliosis, kyphosis or kyphoscoliosis.

INTERACTIVE SUMMARY

As an introduction I have used my Editorial published in the Journal of the Royal Society of Medicine 1982 entitled Management of Scoliosis which outlines the state of knowledge at that time (Paper 1).

A major portion of this work is related to my studies of the pathogenesis, natural history and management of congenital deformities of the spine. These publications (Papers 2 to 13) have been widely accepted internationally as being a major work on this subject and have resulted in invitations to lecture throughout the world. This has included the prestigious Harrington Lecture given at the American Scoliosis Society in 2006 (Paper 12) and the award of the King James IV Professorship from the Royal College of Surgeons Edinburgh in 2001.

Congenital deformities of the spine occur as a consequence of vertebral anomalies developing *in utero* and producing a localized imbalance of the longitudinal growth of the spine. I have classified and described the natural history of these anomalies and have shown that the key to successful treatment depends on three principles. First, an early diagnosis before the child is 5 years old and the curve, whether a scoliosis, kyphosis or kyphoscoliosis exceeds 40 degrees and before the compensatory curves become fixed. Secondly, the ability to anticipate what is likely to happen based on the type and sites of the vertebral anomalies, as well as the age of the patient and the amount of spinal growth remaining. Finally, the application of prophylactic surgical treatment to balance the growth of the spine at an early age and before the development of a significant deformity. In Paper 9 I have described the adverse effect of a congenital thoracic kyphosis or kyphoscoliosis on respiratory function in young children. I have shown that although surgery at a later stage can produce acceptable results (Papers 5 and 7), it is much better to provide prophylactic surgery at an early stage rather than to wait and perform potentially hazardous surgical salvage procedures once a severe rigid deformity has developed.

In Paper 3 I have investigated the incidence of associated intraspinal neural anomalies which if not recognized and treated could have a significant adverse effect on the outcome of surgery to correct the spinal deformity.

Idiopathic scoliosis is a structural lateral curvature of the spine developing without evident clinical or radiological cause in otherwise healthy children. Depending on the age of onset, the scoliosis has been classified into 3 types: infantile (presenting from birth to 3 years), juvenile (presenting from 3 to 10 years) and adolescent (presenting from 10 years to skeletal maturity). There is a genetic tendency to develop these deformities but this, by itself, is insufficient and other factors are necessary to “trigger off” the deformity.

I have shown in Paper 15 that the onset of infantile idiopathic scoliosis is associated with immobility of the new-born causing moulding of the thoracic cage and secondary vertebral rotation producing a coupling effect which results in the development of a lateral curvature of the spine. The majority of these curves will resolve once the infant becomes mobile. However, in a minority, the balance is tipped which results in a rapidly progressive and very severe rigid deformity developing before the age of 10 years. This is a very different condition from that presenting in the juvenile and adolescent years. In Paper 16 I have described the management of these progressive infantile curves. This requires recognition of their poor prognosis at an early stage and the initiation of immediate treatment which necessitates a long and difficult struggle, using both conservative and operative means over many years, to prevent increasing deformity.

The curve patterns and prognosis in juvenile idiopathic scoliosis are described in Paper 18. It was found that juvenile curves are very different from those presenting in the infantile and adolescent years. Infantile scoliosis is more common in boys and the great majority have left-sided thoracic curves. During the juvenile years there is a gradual change in the clinical pattern until, by adolescence, the majority of patients are girls with a high proportion of right-sided thoracic curves. In addition a significant number of infantile curves resolve spontaneously whereas

those that progress can result in a very severe deformity occurring before the age of 10 years. Juvenile curves develop during a period of slow steady spinal growth and unlike infantile progressive curves initially deteriorate relatively slowly. However, after the age of 10 years and during the adolescent growth spurt, there is a rapid progression of the scoliosis and, despite the use of a brace, most patients require spinal surgery in the early teenage years. In contrast, adolescent idiopathic scoliosis develops from a straight spine during the adolescent growth spurt and is therefore subjected to a much shorter period of accelerated spinal growth before stabilizing at skeletal maturity. Only a minority of these adolescent patients require operative treatment. I concluded that because of the difference in prognosis, juvenile curves should continue to be classified as a separate entity.

The effect of the adolescent growth spurt on an early posterior spinal fusion in patients with infantile and juvenile idiopathic scoliosis is described in Paper 19. It was found that a solid posterior fusion will stop the longitudinal growth in the posterior spinal elements but that the vertebral bodies will continue to grow and this becomes most apparent during the period of accelerated growth at puberty. As a result, the vertebral bodies bulge laterally towards the convexity of the scoliosis and pivot on the solid posterior fusion, causing a loss of spinal correction in the frontal plane with increasing vertebral rotation and a recurrence of the rib hump. This publication was the first to report an effect which was later described by other authors as the “crankshaft phenomenon”. At the time of our publication, I suggested that this problem could possibly be overcome by performing an anterior spinal fusion in association with the posterior procedure. This has subsequently become a standard procedure for a scoliosis requiring surgical treatment before the onset of the adolescent growth spurt (Paper 17).

The stability of the scoliotic spine after spinal arthrodesis depends on producing a solid fusion of sufficient strength to resist bending under the influence of gravity and the deforming factors which first caused the curvature. In Papers 21 to 28 I have described the development of my operative techniques for producing a solid stable fusion as well as the evolution of different methods of obtaining

correction of these deformities. In Paper 26, I compared the results of Luque L rod instrumentation using segmental sublaminar wiring with Harrington instrumentation in two matched groups of patients with adolescent idiopathic scoliosis. The use of induced hypotension to control bleeding during posterior arthrodesis has been described (Paper 25) as well as the possible adverse effect of anterior segmental vessel ligation in patients undergoing anterior spinal surgery (Paper 29). The Integrated Spine Imaging System (ISIS) has been used to assess the effect of surgery on the rib hump in adolescent idiopathic patients treated by Luque segmental sublaminar instrumentation (Paper 27).

Spinal deformity occurring as a consequence of neuromuscular disease can present as one of the greatest challenges to the orthopaedic spine surgeon. These patients are very different from those with congenital or idiopathic scoliosis and must be assessed differently in order to avoid serious errors in management. In neuromuscular disease the treatment of the spine is influenced by a number of factors, such as respiratory muscle paralysis, sensory disturbance and functional disability of the limbs which are not present in other types of scoliosis. In Papers 30 to 33 I have shown that spinal surgery for these patients is a complex task but is very rewarding for these severely disabled children.

In Papers 35 to 38 I have described the development and possible problems in the treatment of spinal deformities occurring in association with relatively rare conditions such as Ehlers-Danlos Syndrome, Sotos Syndrome, Di George Syndrome and after cardiac transplantation. The rates of muscle protein synthesis in the paraspinal muscles of patients with adolescent idiopathic scoliosis undergoing surgery has been assessed using stable isotope technology and reported in Paper 39.

In the adult patient, ankylosing spondylitis when allowed to run its full course without treatment can, on occasion, present as one of the most crippling diseases seen by the spine surgeon. The characteristic spinal deformity is a flattening of the normal lumbar lordosis and an increasing thoracic kyphosis with the head and neck thrust forwards and occasionally an increasing kyphosis at the cervico-thoracic

junction. As the disease progresses, the entire spinal column from sacrum to occiput becomes ankylosed by bone in this deformed position. At this stage, the patient is bent forward and forced to look at the ground. This ugly posture is not only functionally disabling but also psychologically disturbing for the patient. In Papers 40 to 43 I have shown that corrective surgical treatment at this late stage to perform a cervical or lumbar spinal osteotomy or a combination of both procedures can be successful in realigning the spine and allowing the patient to see straight ahead. I have described new techniques for performing the osteotomies as well as applying internal fixation in order to correct and stabilize the spine (Paper 41 to 43). Although these are difficult and potentially hazardous procedures successful results can be achieved.

Statement of Originality

I confirm that except for Papers 39 and 40 I was the originator of the idea leading to the research, was responsible for collecting and preparing the material, performing the surgical techniques described and was either solely responsible or the major contributor for all of the work presented.

Where collaboration has taken place, the other authors have in the majority been surgical trainees under my supervision or occasionally more senior colleagues.

All of the papers presented here have been subjected to external referees appointed by the editors of the respective journals in which they were published.

My papers on the Scintigraphic Assessment of the Posterior Fusion Mass which was the subject of my MD thesis (1979) as well as my other publications on spinal problems not related to deformity, have been excluded.

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Section A

Introduction

Editorials

Management of scoliosis

Scoliosis occurring in the growing child is often a progressive condition which can on occasion, if left untreated, result in severe deformity with disability or death from cardiorespiratory failure in adult life (Nilsson & Lundgren 1968, Colles & Ponseti 1969). To prevent these possible complications, all curves should be recognized at an early stage because it is easier to prevent than correct severe deformity.

Scoliosis most frequently presents during the periods of most rapid growth of the spine and all children should, therefore, have their backs examined in the first year of life, on entering school and between the ages of twelve and thirteen years. The prevalence of scoliosis detected by school screening has varied from less than 1% to 13.6% depending on what has been accepted as a significant scoliosis (Wynne-Davies 1968, Brooks *et al.* 1975). Of the curves detected, less than 0.3% will be potentially serious requiring either observation, conservative or operative treatment; the remainder will be non-progressive curves of less than 10°, which are of no significance, and some may improve spontaneously. School screening is best carried out as part of a routine medical examination by an informed school doctor who refers patients to a specialized scoliosis clinic at his own discretion. It is only at this clinic that the scoliosis surgeon can anticipate the prognosis and plan a course of treatment. The general orthopaedic surgeon will never see sufficient children with scoliosis to gain the experience necessary to anticipate the problems or have available the sophisticated technology necessary to treat the condition. At present few children fail to have their curvature recognized at an early stage, but a number still suffer due to the failure of their general practitioner or orthopaedic surgeon to refer them to a scoliosis clinic at an early stage.

If the curve is small and the prognosis uncertain, the condition is best observed by means of regular spinal radiographs which are measured and the rate of progression noted. It is important, however, not to overtreat small non-progressive curves. Should the curve be large when first seen or show signs of progression, treatment is indicated either by conservative or operative means.

Physiotherapy by itself has no part to play in the conservative management of scoliosis. Its use is based on the misguided belief that it is possible

to strengthen unilaterally the muscles on the convexity of the curve, so creating a muscle imbalance which would correct the deformity. This, however, is impossible because exercises strengthen all of the spinal muscles equally. More recent advances using unilateral electrical stimulation of the paraspinal muscles either transcutaneously or by surgically implanted electrodes have been more successful in creating a muscle imbalance. The successful application of this method has so far been limited to small flexible thoracolumbar or lumbar curves of less than 30°. In larger curves the mechanical disadvantage is too great and the curve progresses, although often more slowly. However, this is still an experimental technique and the long-term results are not yet available (Bobechko *et al.* 1979).

Spinal bracing remains the key to the successful conservative management of scoliosis. In the late 1940s Blount and Schmidt of Milwaukee produced a brace used after spinal fusion to correct and stabilize the spine until the fusion became solid (Blount *et al.* 1958). Over the years, however, it was found that the Milwaukee brace, when combined with an exercise programme, was equally effective in the nonoperative treatment of scoliosis as a means of partially correcting and preventing increasing deformity (Moe & Kettelson 1970). The modern Milwaukee brace consists of a carefully moulded plastic pelvic girdle connected to an encircling neck piece by extendable rods which provide a distracting force. The main correcting force is provided, however, by an adjustable pad applied over the rib hump and, when combined with specific exercises, provides the third point in a dynamic three-point correcting system of forces. The major disadvantage of the Milwaukee brace is that, although it is readily accepted by young children, teenage girls understandably dislike the throat piece because of its cosmetic appearance. In order to overcome this problem an underarm type of brace, often known as the Boston brace, has been devised which does away with the throat piece but still relies on the dynamic three-point correction provided by a pressure pad incorporated in the brace (Watts *et al.* 1977). Patients with neuromuscular lesions and reduced sensation are best treated in a total contact underarm brace which spreads the pressure over a wide area, reducing the likelihood of pressure sores.

In selecting the type of brace, any curve whose apex is at or above the eighth thoracic vertebra is best treated in a Milwaukee brace and a curve

below this level can be managed in a Boston brace. There are, however, limitations to the use of both of these braces in that they only work satisfactorily in growing children with relatively flexible curves which are not very severe, i.e. less than 40°. After the brace is applied there is usually an initial improvement of 30–40%, followed by a very gradual loss of correction over a number of years. When the brace is removed at skeletal maturity there is a further slight loss of correction before the curve finally stabilizes at a degree of severity approaching that before the brace was applied. The satisfactory treatment of a patient in a brace is difficult and requires meticulous attention. The best results are obtained when the scoliosis surgeon, orthotist and physiotherapist each see the patient together, and the brace is made in close proximity to the clinic so that immediate alterations can be carried out as required.

The objective of spinal fusion in the treatment of scoliosis is to transform the structural curve into a solid bar of bone which will resist bending under the influence of the factors which caused the deformity. With modern fusion techniques this is a very successful method of preventing deterioration, but there are several important reasons why not all progressive curves are fused. First, one does not wish to perform major spinal surgery with its complications if the scoliosis can be controlled by other means. Secondly, a spinal fusion in a very young child can cause a problem because the fused segment stops growing and by adulthood the trunk will appear very short indeed. Spinal fusion should not, therefore, be performed before the age of ten years unless the scoliosis cannot be controlled by any other means. After the age of ten years the lack of growth in the fused segment becomes much less obvious and a spinal fusion may be performed on any curve with a bad prognosis. Brace treatment is reserved for scoliotic curves with radiographic evidence of progression beyond 25° but still remaining less than 40°. Curves of over 40° may on occasion be treated with braces in young children to allow further growth before spinal fusion, or in major double curves near maturity when we do not wish to fuse the whole spine.

Prior to spinal fusion it is usual to correct the deformity as much as possible without danger to the spinal cord and the correction is maintained by a solid fusion. Preoperative means of correction may involve the use of a Risser-Cotrel type of plaster jacket applied with the curve corrected by traction, halofemoral skeletal traction or Cotrel traction. These methods have the advantage that the curvature is corrected while the patient is

awake and are therefore less likely to produce irreversible neurological complications. They have the disadvantage, however, that the degree of correction is poor in rigid curves and the most effective methods of correction are applied at the time of surgery.

In the 1950s Harrington of Houston developed his technique of posterior spinal instrumentation which has become one of the major contributions to the surgical correction of scoliosis (Harrington 1962). The instruments have remained virtually unchanged and consist of a system of prestressed stainless steel rods and hooks which allow purchase on the posterior bony structures of the spine anywhere from the sacrum to the first thoracic vertebra. The apparatus is in two parts: a distraction system which jacks the curve out and supports the spine by means of a rigid rod applied across the concavity, and a second more flexible rod applied around the convexity and gaining further correction by means of compression. A transverse traction bar has also been devised to connect the two rods and provide even greater stability. Using these techniques it is possible to correct most idiopathic curves by approximately 50–60%.

A rare complication following the sudden distraction of the spine has been the acute onset of lower limb neurological abnormalities thought to be due to stretching of the blood supply to the spinal cord. This complication can on occasion be reversed if the instrumentation is removed within six hours of its insertion. Others have temporarily lightened the anaesthetic after distraction to check the neurological status of the patient before completing the operation (Vauzelle *et al.* 1973, Hall *et al.* 1978). Intraoperative spinal cord monitoring using cortical evoked potentials has also been used, but the readings are difficult to interpret and the method is still experimental (Engler *et al.* 1978).

Harrington at first used his technique as a method of correcting the deformity without fusing the spine or applying external support. Unfortunately, the unrestricted movement caused the metallic hooks to cut out of the bone and it became obvious that spinal fusion with external immobilization was necessary if correction was to be maintained. Recently, Harrington instrumentation without fusion has once more been applied to young children with severe progressive curves in an attempt to gain repeated corrections over a period of years before finally fusing the spine (unpublished). In these children one has to accept a high incidence of instrument failure despite the additional use of external

bracing. The long-term success of Harrington instrumentation, however, depends on the development of a solid stable posterior fusion. Modern posterior fusion techniques, combined with the rigidity provided by Harrington instrumentation, have lowered the incidence of pseudarthrosis to less than 1% in idiopathic scoliosis, and the fusion becomes rapidly stable with only minimal loss of correction after removal of all external support at nine months. The period of bed rest following fusion has in the past been up to six months but it is now possible to mobilize the patient, in an underarm jacket or brace, seven days after surgery and allow a return to all normal activities, except for heavy lifting and contact sports, five weeks after surgery (McMaster 1980). The psychological advantages of decreasing the period of recumbancy and time in hospital are obvious.

The second major advance in the operative correction of severe deformity was devised by Dwyer of Sydney, Australia, in the late 1960s (Dwyer & Schafer 1974). Dwyer initially thought his technique was best applied to idiopathic scoliosis, but the method is now mainly used to correct severe neuropathic curves in patients with myelomeningocele, cerebral palsy and poliomyelitis. The convex side of the scoliosis is approached through an extensive transthoracic and retroperitoneal approach in which the diaphragm is divided circumferentially. The intervertebral discs and vertebral end plates are excised throughout the length of the curve and the spaces packed with bone chips from an excised rib. Starting at one end of the curve, specially designed screw-and-staple units are inserted into each vertebral body and a flexible, braided titanium wire cable is passed through the holes in the heads of the screws. Adjacent vertebrae are compressed and the tension maintained by crimping the screw-heads into the cable. The process is repeated throughout the length of the curve which is gradually straightened. Zielke *et al.* (1978) have modified the technique in an attempt to correct the deformity due to vertebral rotation as well as the lateral curvature. They replaced the Dwyer cable with a solid rod passed through the heads of the screws which are placed to derotate the vertebral bodies as compression is applied.

Using these methods of anterior instrumentation, an excellent degree of correction can be obtained, and the segmental fixation is much more secure than with a Harrington distraction rod which grasps only the two end vertebrae. The risk of traction on the spinal cord is also lessened because the spinal column is shortened rather than lengthened. The major

disadvantages are that for anatomical reasons the screws cannot be easily inserted above the fifth thoracic vertebra or into the sacrum and the incidence of pseudarthrosis is greater than with a posterior fusion. For these reasons anterior instrumentation and fusion are usually combined two weeks later with a posterior fusion and Harrington instrumentation. This allows the fusion to be extended either above or below the anterior fusion if necessary, gives even better correction and produces a very solid and stable fusion. This combination of techniques is best applied to severe thoracolumbar or lumbar scoliosis associated with pelvic obliquity. For the majority of patients who present with either a thoracic scoliosis or a mild-to-moderate thoracolumbar or lumbar scoliosis, posterior fusion with Harrington instrumentation alone has remained the treatment of choice.

The most recent method of spinal instrumentation has been devised by Luque of Mexico. The scoliosis is corrected by segmentally wiring two pre-bent stainless steel rods applied bilaterally to the laminae throughout the curvature (Luque & Cardoso 1977). This gives a very rigid fixation which allows mobilization without external support within a few days of the operation. The method, however, remains experimental and neurological complications can occur when passing the wires beneath the laminae (unpublished).

In the last twenty-five years there have been many major advances in the treatment of scoliosis and there is now virtually no severity of curvature which cannot be significantly improved by surgery. It should be remembered, however, that the necessity for the surgical salvage of severe deformity indicates a failure of management, and the key to successful treatment lies in the early detection and prevention of severe deformity.

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Section B

Congenital Deformities of the Spine

The Natural History of Congenital Scoliosis

A STUDY OF TWO HUNDRED AND FIFTY-ONE PATIENTS

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ABSTRACT: The cases of 251 patients with untreated congenital scoliosis were studied, and 216 patients were followed without treatment for an average of 5.1 years. Most of the curves were first seen when the patient was either in the first two years of life or at puberty, when there was an increased rate of deterioration. An early onset carried a bad prognosis. Of the 143 patients who were last seen, without treatment, after the age of ten years, fifty-one (36 per cent) had a curve of 40 to 60 degrees and forty (28 per cent) had a curve of more than 60 degrees. We concluded that the prognosis in such patients is even worse because sixty-seven patients in our series required treatment at or before the age of ten years due to the severity of the curve: twenty of them had a curve of 40 to 60 degrees and thirty-nine had a curve of more than 60 degrees.

Radiographically, 90 per cent of the curves could be classified into five groups. The rate of deterioration and the ultimate severity of the curve were found to depend on both the type of anomaly and the site at which it occurred. Increasingly severe and progressive scoliosis, regardless of the area of the spine affected, developed when there was a block vertebra, a wedge vertebra, a single hemivertebra, two unilateral hemivertebrae, a unilateral unsegmented bar, or, most severe, a unilateral unsegmented bar with contralateral hemivertebrae at the same level. For each of these types of anomaly, the rate of deterioration was usually less severe if the abnormality was in the upper thoracic region, more severe in the thoracic region, and most severe in the thoracolumbar region. The median yearly rate of deterioration for each type and site of curve without treatment before and after the patient was ten years old was evaluated. Secondary problems due to tilting of the head, elevation of the shoulder line, decompensation of the trunk, pelvic obliquity, and the formation of large secondary structural curves also occurred, and contributed significantly to the over-all disability and deformity.

Congenital scoliosis is a lateral curve of the spine that is due to the presence of vertebral anomalies that cause an

imbalance in the longitudinal growth of the spine. These vertebral anomalies develop during the first six weeks of intrauterine life, when the anatomical pattern of the spine is formed in mesenchyme. Once the mesenchymal mold is established, the cartilaginous and osseous stages follow that pattern. The vertebral abnormality is present at birth, but the clinical deformity may not become evident until later in childhood when a scoliosis develops and the diagnosis can be made radiographically. Some anomalies cause so little deformity that they remain undetected, so the true incidence of congenital scoliosis in the general population remains unknown. Wynne-Davies found that multiple vertebral anomalies were often hereditary in origin, but the occurrence of an isolated single anomaly was usually sporadic.

The radiographic appearance of the vertebral anomalies varies considerably and, as a result, congenital scoliosis was for many years thought to be unpredictable in its behavior. Some thought that it seldom required treatment^{1,3}. In 1952, Kuhns and Hormell reviewed the cases of 165 children and concluded, as did many physicians at that time, that a congenital scoliosis usually is relatively benign and progresses slowly, if at all. It was not until 1968 that Winter et al., in a study involving 234 children, firmly established the much more serious prognosis for certain types of congenital scoliosis. Apart from these two papers, there have been very few reports of the natural history of congenital scoliosis in large numbers of unselected patients⁶.

A congenital scoliosis often is rigid and correction can be difficult. It therefore is important to be able to anticipate when a congenital scoliosis is at risk for rapid deterioration and to initiate treatment when the curve is small rather than to attempt the dangerous surgical salvage procedures that are necessary when the deformity is severe. Planning such a prophylactic course of treatment requires a more thorough knowledge of the natural history of all types of congenital scoliosis than is presently available.

The purpose of this study was to investigate the natural history of congenital scoliosis in a large number of patients. All came from one area and were seen at the Edinburgh Scoliosis Clinic, and most were followed for long periods. This clinic is the only referral point for patients with congenital scoliosis from a large population. It was hoped that this paper would provide indications as to the necessity for treatment of the various types of congenital scoliosis.

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Classification

We classified the types of vertebral anomaly causing congenital scoliosis in the patients in this study according to the system of Winter et al. We added one subcategory, as described by Nasca et al., because it has a particularly severe prognosis. Under the category of simple anomalies, we included failure of segmentation, which consists of: (1) a unilateral unsegmented bar, (2) unilateral failure of segmentation with contralateral hemivertebrae at the same level⁶, and (3) bilateral failure of segmentation (block vertebrae).

The other simple anomaly in our series was unilateral failure of formation of a vertebral body, which was either: (1) complete (hemivertebra) or (2) partial (wedge vertebra).

Complex (unclassifiable) anomalies consisted of a jumble of vertebral anomalies that were too varied for separate classification. In some patients included in this group it was impossible to identify the type of anomaly because the severity of the curve was so great.

Occasionally other congenital vertebral anomalies were present at levels in the spine other than those involving the scoliotic curve, but if they did not contribute to the scoliosis they were ignored in the classification. Anomalies of the ribs were very common, but they too were ignored because they did not cause a scoliosis by themselves.

In addition to determining the type of vertebral anomaly, the scoliosis was also classified according to the site of the apex of the curve, which frequently coincided with the site of the anomaly. A scoliosis whose apex lay between the second and sixth thoracic vertebrae, inclusive, was termed an *upper thoracic curve*. A scoliosis whose apex was located between the seventh and eleventh thoracic vertebrae, inclusive, was a *lower thoracic curve*. A *thoracolumbar scoliosis* was one with its apex at the twelfth thoracic or first lumbar vertebra. The apex of a *lumbar scoliosis* was between the second and fourth lumbar vertebrae, inclusive, and the apex of a *lumbosacral scoliosis* was at the fifth lumbar vertebra⁸.

A patient with a single congenital scoliosis was considered to have only one structural curve due to the presence of congenital vertebral anomalies. Several patients were classified as having a single congenital scoliosis even though a second structural scoliosis developed either above or below the primary congenital curve. This second curve was not classified as a congenital scoliosis because it did not result from a congenital vertebral anomaly.

A patient was considered to have a double congenital scoliosis if there were two opposing structural curves, each due to separate congenital vertebral anomalies that might or might not be similar. A few patients had more than two structural curves because of separate congenital vertebral anomalies.

Skeletal maturity was determined radiographically when there was complete ossification and fusion of the iliac apophyses¹¹.

Materials and Methods

A study was made of all patients seen at the Edinburgh Scoliosis Clinic between 1958 and 1981 who were diagnosed as having congenital scoliosis. The majority of these patients were seen after 1965. Patients who had myelomeningocele, Klippel-Feil syndrome, or congenital kyphosis or kyphoscoliosis in which the kyphosis was the predominant deformity were excluded because the natural history of those deformities was very different from that of the ones under study. There were 251 patients who had a true congenital scoliosis that was diagnosed radiographically; that is, each had a lateral curve of the spine due to the presence of congenital vertebral anomalies. Radiographs made elsewhere, prior to referral, were often available, and sometimes provided evidence of progression of the deformity.

There were 179 female and seventy-two male patients, whose ages at diagnosis ranged from birth to twenty-two years (Fig. 1). They had a total of 269 curves (Table I).

Two hundred and sixteen patients were followed for a mean of 5.1 years (range, six months to seventeen years). At their last visit to the clinic, 108 patients had not been treated, and forty-six of them had reached skeletal maturity. The mean length of follow-up for these patients was 7.3 years. The sixty-two patients who were skeletally immature were still attending our clinic, without treatment, after a mean of 5.1 years. The remaining 108 patients had been followed without treatment for a mean of 4.2 years before eventually being treated either with a Milwaukee brace or with spine fusion; the data on these patients relate to the pretreatment interval.

No longitudinal data could be obtained on the natural history of the remaining thirty-five patients, since twenty-three of them received immediate treatment when they were first seen because of the severity of the deformity and the other twelve were skeletally mature when first seen. The findings in these patients are included in some of the analyses in this study because prior to attending our clinic they had been untreated, and therefore contributed data on the natural history of congenital scoliosis.

The sites and types of congenital scoliosis were diagnosed by reviewing the radiographs of the spine, made with the patient standing. Using the Cobb method, care was taken to remeasure from the same levels that were used on the earliest radiograph. Pelvic obliquity was measured on the radiographs of the spine by noting the angle subtended between a line drawn across the tops of the iliac crests and the bottom of the radiograph, which was horizontal when the patient was standing. Decomposition or listing of the trunk and tilting of the head were assessed on the basis of the spine radiographs and the clinical photographs made with the patient standing.

Results

The incidences of the various sites and types of congenital scoliosis are shown in Table I.

TABLE I
SITES AND TYPES OF CONGENITAL SCLIOSIS (TWO HUNDRED AND FIFTY-ONE PATIENTS)

Site of Curve	Type of Congenital Vertebral Anomaly (No. of Curves)					
	Unilateral Unsegmented Bar	Unilateral Unsegmented Bar with Contralateral Hemivertebrae	Block Vertebra	Hemivertebra	Wedge Vertebra	Unclassifiable
Upper thoracic	40	7	7	23	1	5
Lower thoracic	39	11	2	21	4	13
Thoracolumbar	17	10	3	13	3	8
Lumbar	6	0	1	19	1	3
Lumbosacral	0	0	0	12	0	0

A single congenital curve occurred in 238 patients: 216 of the curves were due to simple anomalies and twenty-two, to complex anomalies.

Two or more congenital curves occurred in thirteen patients. In ten of the patients the curves were due to simple anomalies and in three the anomalies were complex.

The ages of the patients and the sizes of the congenital curves at the initial visit are shown in Figure 1. In patients with two or more congenital curves, only the largest curve was used for tabulation. Most of our patients were diagnosed radiographically either in the first two years of life (seventy-seven patients) or between the ages of nine and fourteen years (sixty-eight patients). The degree of severity of the curve did not correlate with the age at presentation. Severe curves of more than 80 degrees were

seen not only in older children but also at birth and in the first year of life. Minor curves were seen in patients in all age groups.

Severity of the Curve and Rate of Deterioration

Of the 251 patients in this study, 173 were seen, untreated, at or before the age of ten years. Thirty-eight patients (22 per cent) had a curve of 20 degrees or less, forty-seven (27 per cent) had a curve of 21 to 39 degrees, forty-one (24 per cent) had a curve of 40 to 60 degrees, and forty-seven (27 per cent) had a curve of more than 60 degrees (range, 62 to 148 degrees).

Sixty-seven of these 173 patients were eventually treated at or before the age of ten years because of the severity of the deformity. Before treatment, eight had a

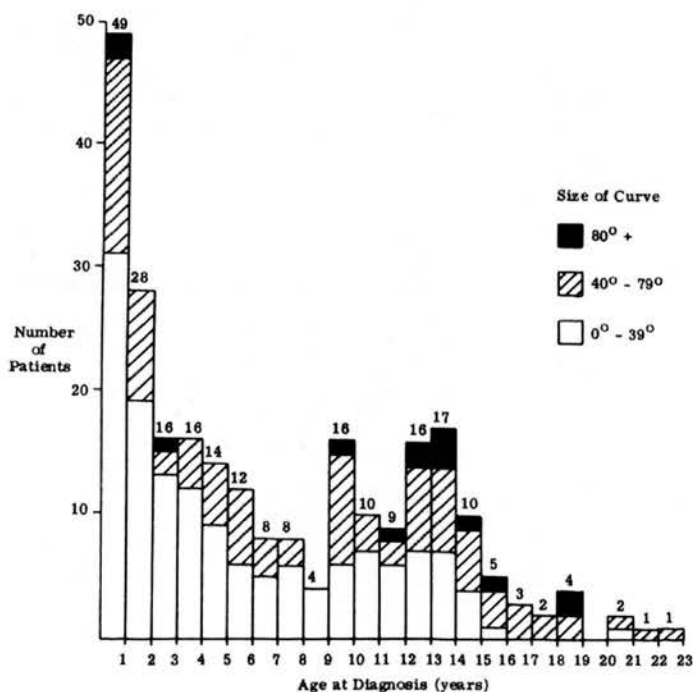


FIG. 1

Ages of the patients and severity of the congenital scolioses at diagnosis.

curve of 20 to 39 degrees, twenty had a curve of 40 to 60 degrees, and thirty-nine had a curve of more than 60 degrees.

One hundred and forty-three patients were last seen, without treatment, after they were ten years old. Fifteen of them (10 per cent) had a curve of 20 degrees or less, thirty-seven (26 per cent) had a curve of 21 to 39 degrees, fifty-one (36 per cent) had a curve of 40 to 60 degrees, and forty (28 per cent) had a curve of more than 60 degrees (range, 64 to 156 degrees).

Fifty-eight patients reached skeletal maturity without treatment. Of these patients, six (10 per cent) had a curve of 20 degrees or less, twenty-two (38 per cent) had a curve of 21 to 39 degrees, nineteen (33 per cent) had a curve of 40 to 60 degrees, and eleven (19 per cent) had a curve of more than 60 degrees (range, 66 to 156 degrees).

All of the large curves that were followed without treatment should have been treated at a much earlier stage, regardless of the patient's age.

We found, as did Winter et al., that the rate of deterioration and the ultimate severity of a congenital scoliosis depended on both the type of vertebral anomaly and the site of the curve. In addition, we found that the rate of deterioration was not constant but tended to increase after the age of ten years.

In order to obtain a more complete understanding of the natural history of all types of congenital scoliosis, we studied the rate of deterioration in degrees per year and the severity of the curve, both before and after the age of ten years, for each type of vertebral anomaly producing a scoliosis in each region of the spine (Tables II through VI). The degree of scoliosis was followed serially and remained fairly constant until between the ages of nine and eleven years, when it tended to increase, especially in patients with unilateral failure of segmentation. The rate of deterioration before and after the age of ten years was estimated by subtracting the initial angle from the final angle of the curve on the routine spine radiographs made with the patient standing, and dividing the increment by the number of years and months. We calculated the median rate of deterioration rather than the mean for each specific type of congenital scoliosis occurring at each specific site, and this value was found to be more representative of each subgroup as a whole and less distorted by values that were greatly different from those of the majority (Fig. 8).

Multiple Congenital Curves

Before analyzing the single curves, we studied the cases of the thirteen patients with two or more congenital curves due to simple (Table II) and complex anomalies.

Three patients (Cases 1, 2, and 3) had double curves due to unilateral unsegmented bars on opposite sides and in different regions of the spine. These double curves showed a high rate of deterioration and became large and unbalanced, causing a list of the trunk that necessitated correction and fusion soon after the patient was ten years old.

Four patients had two hemivertebrae, but on opposite sides and at different levels of the spine. In two of these patients (Cases 4 and 5) the hemivertebrae occurred in the thoracic region, within one or two segments of each other and on opposite sides of the spine. These hemivertebrae caused two small kinks that never became large and that balanced each other, producing a minimum deformity at maturity. Two other patients (Cases 6 and 7) had opposing hemivertebrae that were much more widely separated, on opposite sides, and in different regions of the spine. These hemivertebrae produced bigger curves that were unbalanced, causing a list of the trunk and requiring correction and fusion at the ages of ten and fourteen years.

Two patients (Cases 8 and 10) had three hemivertebrae and one patient (Case 9) had four hemivertebrae which alternated on either side of the spine in the thoracic and thoracolumbar regions. The resulting curves tended to balance each other, but in one patient (Case 8) the upper hemivertebra was at the first thoracic level and this caused an elevation of the shoulder line, requiring correction and fusion of the upper curve at the age of eight years.

Three patients had multiple small curves due to complex anomalies. These curves were closely associated with one another and tended to balance each other, causing little deformity other than stunting of the spine. One patient, however, had an elevated shoulder line that necessitated correction and fusion of the upper thoracic curve at the age of nine years.

Single Congenital Curves

For the purpose of analysis of the natural history of the curves, each of the six groups was divided into three subgroups. Subgroup A comprised those patients for whom serial observations had not been made because they were either treated immediately or were first seen, untreated, at skeletal maturity. Subgroup B contained those patients who had been followed for variable periods before the age of ten years. Subgroup C contained those patients who had been followed at or after the age of ten years. There was some overlap between Subgroups B and C.

Unilateral Failure of Segmentation (Unilateral Unsegmented Bar) (Group 1)

A unilateral unsegmented bar was the cause of a congenital scoliosis in ninety-nine patients and was always present on the concavity of the curve. Ninety-six patients had a single congenital scoliosis and three (Table II, Cases 1, 2, and 3) had two separate and opposing unbalanced congenital curves, each due to a unilateral unsegmented bar but on opposite sides and at different levels of the spine. The unsegmented bars in the ninety-nine patients did not demonstrate a preference for side but were seen more frequently in the thoracic region (seventy-nine patients) than elsewhere. In three patients the unilateral unsegmented bar was not recognized in infancy and was only diagnosed retrospectively, between the ages of three and four years, when the bar began to ossify and appeared on

TABLE
NATURAL HISTORY OF TEN PATIENTS WITH MORE

Case	Sex	Vertebral Anomaly		Curve		Age (Yrs. + Mos.)		Length of Follow-up (Yrs. + Mos.)
		Type	Site	Side	Extent	When First Seen	When Last Seen	
1	F	Unseg. bar	T4-T9	L	T3-T12	3 + 0	13 + 6	10 + 6
		Unseg. bar	L2-L4	R	L1-L5			
2	F	Unseg. bar	T2-T5	L	T2-T5	0 + 2	2 + 3	2 + 1
		Unseg. bar	T11-L1	R	T11-L3			
3	F	Unseg. bar	T3-T5	R	T1-T7	8 + 2	11 + 0	1 + 8
		Unseg. bar	L1-L3	L	T7-L3			
4	F	Hemivert.	T3	L	T2-T4	2 + 3	17 + 0	14 + 9
		Hemivert.	T6	R	T5-T7			
5	F	Hemivert.	T2	R	T1-T3	3 + 1	4 + 4	1 + 3
		Hemivert.	T5	L	T4-T6			
6	F	Hemivert.	T7	L	T2-T12	3 + 0	10 + 0	7 + 0
		Hemivert.	L2	R	T12-L5			
7	F	Hemivert.	L1	L	T11-L4	13 + 11		None
		Hemivert.	L6	R	L4-S1			
8	M	Hemivert.	T1	R	C7-T3	6 + 3	8 + 1	1 + 10
		Hemivert.	T10	R	T8-T11			
		Hemivert.	L1	L	T11-L4			
9	F	Hemivert.	T6	L	T4-T8	20 + 0		None
		Hemivert.	T9	R	T8-T11			
		Hemivert.	T12	L	T11-L1			
		Hemivert.	L4	R	L3-L5			
10	F	Hemivert.	T5	L	T3-T6	0 + 5	3 + 6	3 + 0
		Hemivert.	T8	R	T7-T9			
		Hemivert.	L4	L	L1-L5			

radiographs. The rate of progression of these three curves did not differ significantly from that of the other curves in the same regions. The prognosis for congenital scoliosis due to a unilateral unsegmented bar depended mainly on the site of the curve and partly on the extent of the bar. The extent ranged from two to eight vertebrae. However, there was not always a direct relationship between the length of the bar and the rate of progression of the curve. The longer unsegmented bars all tended to produce the larger curves in a specific region, but occasionally a shorter bar produced an equally large curve in the same region.

The seventy-two patients who were followed without treatment were considered to be in Groups 1B and 1C (Table III). The twenty-four patients for whom serial observations had not been made were in Group 1A and were excluded from further analysis.

Upper Thoracic Curves

Of the thirty-eight patients who had an upper thoracic curve, ten were in Group 1A. Nine of them were treated immediately when they were first seen, between the ages of two and fifteen years, and had curves ranging from 26 to 85 degrees. One patient was seen at skeletal maturity, with a curve of 68 degrees.

The remaining twenty-eight patients with an upper thoracic curve were in Groups 1B and 1C (Table III) and were followed without treatment for a mean of 4.7 years (range, nine months to fourteen years and two months). Before they reached the age of ten years, the median rate of deterioration was 2 degrees per year. By the age of ten

years, ten of them had a curve of more than 30 degrees. Twelve patients were seen, untreated, at the age of ten years, when the mean curve was 33 degrees (range, 13 to 76 degrees). Two patients had a more rapid rate of deterioration before the age of ten years. One of them, who had a bar involving seven vertebrae, had a 5-degree curve at birth which deteriorated at a rate of 6 degrees per year until it was 62 degrees at the age of ten years (Figs. 3-A and 3-B). The second patient had a bar involving three vertebrae which produced a 24-degree curve at the age of three years and deteriorated at a rate of 5 degrees per year until it was 53 degrees at the age of nine years. The case of this patient demonstrated that even a relatively short unsegmented bar can produce a large curve.

After the patients were ten years old, the median rate of deterioration increased to 4 degrees per year, and sixteen patients required a spine fusion at a mean age of thirteen years (range, eleven to fifteen years), at which time the mean curve was 59 degrees (range, 30 to 98 degrees). The most rapid rates of deterioration occurred in four of these patients, who were followed untreated prior to spine fusion for periods ranging from nine months to two years and six months. A bar involving three to five vertebrae was present in three of these curves, which deteriorated at a rate of 8 degrees per year. A bar involving seven vertebrae was present in one curve, which deteriorated at a rate of more than 10 degrees per year and measured 90 degrees at the age of twelve. Not all of the upper thoracic curves deteriorated at the same rate, however, and five of the ten curves that were followed, untreated, to maturity then

II

THAN ONE CURVE DUE TO SIMPLE ANOMALIES

When First Seen	Size of Curve (Degrees)		Rate of Change per Year	Comments
	When Last Seen	Total Change		
22	122	100	9.5	Unbalanced; upper curve corrected and fused at 13 yrs. and 6 mos. old
0	75	75	7	
8	20	12	6	Unbalanced; Milwaukee brace applied at 2 yrs. and 3 mos. old
7	40	33	16	
25	40	15	9	Unbalanced; required correction and fusion at 11 yrs. old
32	53	21	13	
22	45	23	1.5	Balanced
33	52	19	1	
28	29	1	1	Balanced
27	28	1	1	
39	54	15	2	Unbalanced; required correction and fusion at 10 yrs. old
44	61	17	2	
76				Unbalanced; required correction and fusion at 13 yrs. and 11 mos. old
42				
34	34	0	0	High shoulder and upper curve required correction and fusion at 8 yrs. and 1 mo. old
35	35	0	0	
41	45	4	2	
32				Balanced and skeletally mature
38				
32				
17				
16	16	0	0	Balanced
15	16	1	0	
18	18	0	0	

measured 30 to 40 degrees. The largest untreated curves at maturity measured 68 and 88 degrees and were due to bars involving three and five vertebrae, respectively.

Upper thoracic curves, especially those whose apex lay at the second, third, or fourth thoracic vertebra, produced a significant cosmetic deformity due to elevation of the shoulder line on the convex side of the curve. Tilting of the head toward the concavity also occurred in those patients whose curves extended beyond the cervicothoracic junction; the tilting resulted because a satisfactory compensatory curve failed to develop above the congenital curve (Figs. 2-A and 2-B).

In thirteen patients with an upper thoracic curve, with the apex at the fourth, fifth, or sixth thoracic vertebra, an additional long structural curve developed in the lower thoracic or thoracolumbar region (Figs. 3-A and 3-B). Why such a curve should develop in some patients and not in others is unknown, as it was not always associated with the larger upper thoracic curves. This initially compensatory curve, which involved no congenital anomalies, appeared secondarily; while it initially was compensatory and therefore could be corrected, later it became fixed and deteriorated even more rapidly, and it was soon more severe than the primary curve. The major deformity then was caused by the secondary structural curve, which was much more rotated than the primary congenital curve and produced a large rib hump. Five of the patients with this condition were in Group 1A; two were skeletally mature and three required immediate treatment of the secondary structural curve between the ages of fourteen and sixteen years.

The remaining eight patients were followed without treatment for a mean of 4.8 years (range, nine months to fourteen years). The median rate of deterioration for the five patients followed before the age of ten years was 6 degrees per year (range, 2 to 8 degrees) for the secondary structural curves and 3 degrees per year (range, 1 to 4 degrees) for the primary congenital curves. Five patients were followed after the age of ten years and then the median rate of deterioration was 7 degrees per year (range, 3 to 10 degrees) for the secondary structural curves and 4 degrees per year (range, 2 to 8 degrees) for the primary congenital curves. The structural compensatory curve usually deteriorated at approximately twice the rate of the primary congenital curve. Six patients were treated between the ages of twelve and sixteen years, when the upper thoracic congenital curves were a mean of 56 degrees (range, 30 to 85 degrees) and the lower secondary structural curves were a mean of 79 degrees (range, 53 to 104 degrees). Three patients reached skeletal maturity without treatment, at which time the upper thoracic congenital curves were 40, 59, and 68 degrees and the lower secondary structural curves were 78, 75, and 82 degrees, respectively.

Lower Thoracic Curves

Of the thirty-eight patients who had a lower thoracic curve, nine were in Group 1A. Six of them were treated immediately, between the ages of eleven and fourteen years, with curves ranging from 41 to 96 degrees (mean, 62 degrees). Three were seen at skeletal maturity, with curves measuring from 66 to 89 degrees.

TABLE
UNTREATED SINGLE CONGENITAL SCOLIOSIS DUE

Site of Curve	No. of Curves	No.	Age When First Seen* (Yrs.)	Size of Curve (Degrees)*		Rate of Deterioration per Year (Degrees)									
				When First Seen	When Last Seen before 10 Yrs. Old										
						1	2	3	4	5	6	7	8	9	10
Upper thoracic	28	15	4 (0-9)	29 (0-60)	35 (13-76)	6	3	4	1	1					
Lower thoracic	29	21	2 (0-9)	40 (12-60)	61 (25-84)	3	3	2	1	2	1	5	4		
Thoracolumbar	12	11	3 (0-9)	35 (15-75)	58 (35-82)		1	1		3	5	1			
Lumbar	3	2	(0-1)	(8-62)	(53-82)					1					1

* Median, with range in parentheses.

† The number of patients who were followed untreated until after the age of ten years but who were first seen before the age of ten years, and who are also included in Group 1B, is in parentheses.

The remaining twenty-nine patients with a lower thoracic curve were in Groups 1B and 1C (Table III) and were followed without treatment for a mean of 3.7 years (range, six months to eleven years and seven months). Before the patients reached the age of ten years, many of the curves deteriorated rapidly (at a median rate of 5 degrees

per year) and nine curves deteriorated at a rate of 7 or 8 degrees per year. Ten patients received treatment before the age of ten years. By the age of ten, sixteen of twenty-one curves were more than 40 degrees, and twelve of these were greater than 60 degrees (Figs. 4-A and 4-B). Ten patients were seen, untreated, at the age of ten years, at



FIG. 2-A



FIG. 2-B

A girl, thirteen years old, who had a unilateral unsegmented bar on the right, extending from the second to the fifth thoracic vertebra and producing a right upper thoracic scoliosis measuring 60 degrees. She had a major cosmetic deformity due to elevation of the left shoulder line and slight tilting of the head to the right.

III

TO A UNILATERAL UNSEGMENTED BAR

Group 1C (Patients Seen at or after the Age of Ten Years)											
No.†	Size of Curve (Degrees)*		Age When Last Seen* (Yrs.)	Rate of Deterioration per Year (Degrees)							
	When First Seen at or after 10 Yrs. Old	When Last Seen or at Maturity		2	3	4	5	6	7	8	9
20 (7)	32 (17-81)	47 (20-98)	15 (11-19)	5	2	5	2	2		3	1
14 (6)	43 (25-73)	65 (33-96)	14 (11-18)	1	4	1	1	1			6
6 (5)	58 (35-80)	78 (45-93)	12 (11-17)						1	2	3
1 (0)	20	37	14				1				

which time the mean curve was 48 degrees (range, 25 to 68 degrees). Three of these curves were fused when the patients were ten years old.

After the patients were ten years old, the median rate of deterioration increased to 6.5 degrees per year. Ten patients received treatment at a mean age of thirteen years (range, eleven to sixteen years), at which time the mean curve was 65 degrees (range, 40 to 96 degrees). Three patients reached skeletal maturity without treatment; at that time the curves were 47, 60, and 66 degrees.

The most extensive unsegmented bars in this region occurred in three patients and involved five to six vertebrae. In one of these patients a 45-degree curve was diag-

nosed at birth which, without treatment, became 77 degrees at the age of four years. The other two patients both were first seen at the age of fourteen years with curves of 57 and 50 degrees, which deteriorated rapidly to 70 and 96 degrees in the patients' fifteenth and sixteenth years, respectively.

Thoracolumbar Curves

Of the fifteen patients who had a thoracolumbar curve, three were in Group 1A. Two of them were treated immediately, at two and thirteen years old, with curves measuring 35 and 85 degrees, respectively. One was seen at skeletal maturity, with a curve of 81 degrees.

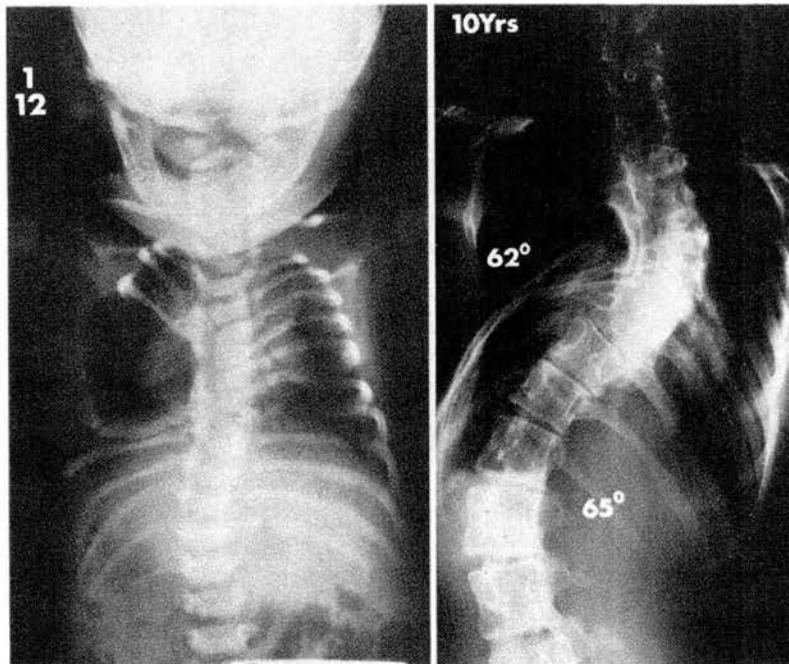


FIG. 3-A

FIG. 3-B

Fig. 3-A: A girl, one month old, who had a radiograph made because of asymmetry of the thoracic rib cage. The radiograph shows absent ribs and a unilateral unsegmented bar extending from the third to the seventh thoracic vertebra on the left.

Fig. 3-B: No treatment was given, and by the age of ten years the unilateral unsegmented bar had produced a 62-degree right upper thoracic scoliosis. The major deformity, however, was due to a very severe secondary left structural thoracolumbar scoliosis that involved no congenital anomalies.

The remaining twelve patients with a thoracolumbar curve were in Groups 1B and 1C (Table III); they were all seen in the first decade of life and were followed without treatment for a mean of five years (range, six months to sixteen years). Before they reached the age of ten years, the median rate of deterioration of the curves was 6 degrees per year. Six patients were treated between the ages of two and seven years, at which time the mean curve was 64 degrees (range, 40 to 82 degrees). The remaining six curves had a mean measurement of 59 degrees (range, 35 to 80 degrees) at the age of ten years.

After the patients were ten years old, the median rate of deterioration increased to 9 degrees per year. Five of six patients received treatment at a mean age of 11.8 years (range, eleven to twelve years), at which time the mean curve was 78 degrees (range, 45 to 90 degrees).

The most extensive bars in this region occurred in three patients and involved six to eight vertebrae. In one patient a 15-degree curve was diagnosed in the first year of life, and at skeletal maturity the untreated curve measured 93 degrees. The second patient was first seen and treated at thirteen years old with an 85-degree curve, and the third was first seen at skeletal maturity with an 80-degree curve.

Pelvic obliquity or listing of the trunk to the side of the unsegmented bar, or both, occurred in twelve of the fifteen patients with a thoracolumbar curve. The degree of pelvic obliquity was severe (more than 25 degrees) in four (range, 26 to 30 degrees), moderate (10 to 25 degrees) in four, mild (less than 10 degrees) in two, and absent in five. The listing was severe in one, moderate in two, mild in five, and absent in seven.

Lumbar Curves

Of the five patients with a lumbar curve, two were treated immediately and were in Group 1A. These two patients had bars involving four and eight vertebrae, producing curves of 64 and 100 degrees at the ages of sixteen and nine years, respectively.

The remaining three patients with a lumbar curve were in Groups 1B and 1C (Table III) and were followed without treatment for a mean of four years (range, one to eight years). One patient had a bar involving three vertebrae which produced a 62 degree curve at the age of one year; the curve had deteriorated to 82 degrees in the second year of life, when it was treated. The second patient also had a bar involving three vertebrae but this produced only an 8-degree curve in the first year of life; the curve deteriorated to 53 degrees at the age of ten years. The third patient had a bar that involved two vertebrae and the curve deteriorated slowly. The curve measured 20 degrees when the patient was eleven years old and then deteriorated more rapidly, to reach 37 degrees when the patient was fourteen years old.

Pelvic obliquity and listing of the trunk occurred in all five of the patients in this group and was directly proportional to the severity of the curve. In the larger curves the pelvic obliquity was the major deforming factor. The se-

vere disability was then due to an apparent shortening of one lower limb.

Unilateral Failure of Segmentation (Unilateral Unsegmented Bar with Contralateral Hemivertebrae) (Group 2)

Twenty-eight patients had a single congenital scoliosis due to a unilateral unsegmented bar on the concavity of the curve with one or more hemivertebrae on the convexity at the same level. Twelve curves were convex to the right and sixteen, to the left. These curves were classified separately from those that were due to a unilateral unsegmented bar alone because they had an even more severe prognosis. By the age of five years, all of these curves became very severe. The rate of deterioration depended mainly on the site of the anomalies and partly on the extent of the unsegmented bar. The bar involved from two to eight vertebrae. The number of hemivertebrae ranged from one to seven. This type of anomaly occurred most frequently in the thoracic region (eighteen patients). The hemivertebrae were most often opposite the bar and not separated by a number of normal vertebrae, as was usually the case when two unilateral hemivertebrae occurred in the absence of a bar. The anomaly was most easily recognized radiographically in the first few years of life (Fig. 5A), but as the curve rapidly increased the hemivertebrae became obscured (Fig. 5B) and were difficult to distinguish from a unilateral unsegmented bar alone. The three patients for whom serial observations were not made were in Group 2A and were not included in this part of the analysis. The twenty-five patients who were followed without treatment were in Group 2B (Table IV).

Upper Thoracic Curves

The seven patients with an upper thoracic curve were in Group 2B (Table IV) and were followed without treatment for a mean of 4.6 years (range, six months to twelve years). Before they reached the age of ten years, the median rate of deterioration of the curves was 5 degrees per year (range, 3 to 8 degrees). Four patients were treated between the ages of three and five years. One of these patients, with an unsegmented bar involving eight vertebrae with seven contralateral hemivertebrae, had an 82 degree curve at birth which deteriorated at a rate of 8 degrees per year to become 108 degrees at the age of four years. The other three patients had unsegmented bars whose extent ranged from three to five vertebrae with one or two contralateral hemivertebrae, and the curves measured 35, 52, and 54 degrees prior to treatment. One patient with a bar involving three vertebrae and one contralateral hemivertebra had a 27 degree curve at the age of three months, and was untreated at the age of two years when the curve had increased to 35 degrees.

Two patients reached the age of ten years without treatment. One, with a bar involving six vertebrae with two contralateral hemivertebrae, had a 45-degree curve at one year old, which deteriorated at a rate of 5 degrees per

TABLE IV
UNTREATED SINGLE CONGENITAL SCOLIOSIS DUE TO A UNILATERAL
UNSEGMENTED BAR WITH CONTRALATERAL HEMIVERTebrae

Site of Curve	No. of Curves	No.	Age when First Seen* (Yrs.)	Size of Curve (Degrees)*		Rate of Deterioration per Year (Degrees)											
				When First Seen	When Last Seen before 10 Yrs. Old												
						1	2	3	4	5	6	7	8	9	10	>10	
Upper thoracic	7	7	1 (0-5)	45 (12-82)	54 (35-108)			1	1	3			2				
Lower thoracic	10	10	1 (0-6)	58 (23-76)	84 (27-108)				1	1	5				1	2	
Thoracolumbar	8	8	1 (0-2)	58 (41-86)	79 (54-148)						1	1		1		5	

* Median, with range in parentheses.

year to become 83 degrees by the age of ten years. The second patient, with a bar involving three vertebrae with one contralateral hemivertebra, had a 40-degree curve at the age of five years old, which deteriorated at a rate of 4 degrees per year to become 59 degrees at the age of ten years. After these patients reached ten years old, the rate of deterioration increased to 5 and 7 degrees per year, and both patients were treated with spine fusion at the ages of eleven and thirteen years old when the curves were 88 and 82 degrees, respectively.

Elevation of the shoulder line or tilting of the head, or both, occurred in all of these patients and, as with other types of upper thoracic congenital scoliosis, caused a significant cosmetic deformity. In addition, in four patients in whom the apex of the curve was at the fourth, fifth, or sixth thoracic vertebra a secondary structural lower thoracic or thoracolumbar scoliosis developed, but did not result from congenital anomalies. These curves deterior-

ated at a rate of more than 5 degrees per year and measured between 59 and 103 degrees in the three patients who were treated between the ages of three and five years. In one patient, who was treated at the age of thirteen years, the curve measured 72 degrees.

Lower Thoracic Curves

Of the eleven patients who had a lower thoracic curve, one was in Group 2A. This patient, with a 60-degree curve, was treated immediately at the age of ten years.

The remaining ten patients with a lower thoracic curve were in Group 2B (Table IV), and were followed without treatment for a mean of 5.5 years (range, one year to ten years and six months). Because of the severity of the deformity, in eight patients the scoliosis was diagnosed in the first two years of life when the mean curve was 50 degrees (range, 23 to 76 degrees). After the initial diagnosis,



FIG. 4-A

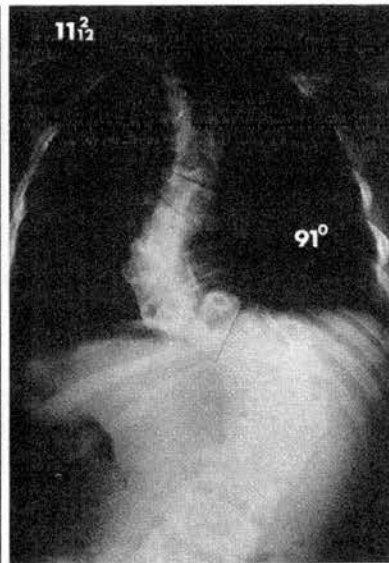


FIG. 4-B

Fig. 4-A: A girl, five years and one month old, with a unilateral unsegmented bar on the right, extending from the eighth to the tenth thoracic vertebra and producing a left lower thoracic scoliosis measuring 61 degrees.

Fig. 4-B: No treatment was given, and by the age of eleven years and two months the lower thoracic curve had deteriorated to 91 degrees.

the median rate of deterioration, without treatment, was 6 degrees per year (range, 4 to 11 degrees) and all but one of the patients received treatment before the age of ten years. Three patients were treated at the age of nine years, with curves of 72, 99, and 106 degrees.

The most extensive vertebral anomalies in this region occurred in four patients who had unsegmented bars involving from seven to nine vertebrae with three to six contralateral hemivertebrae. When the patients were three years old, all of these curves measured more than 70 degrees (range, 76 to 102 degrees). One patient was followed without treatment and at the age of ten years the curve measured 108 degrees. The rate of deterioration then increased, and when the patient was thirteen years old the curve measured 130 degrees.

Thoracolumbar Curves

Of the ten patients who had a thoracolumbar curve, two were in Group 2A. Both of these patients required treatment in the first year of life, with curves of 68 and 75 degrees.

The remaining eight patients with a thoracolumbar curve were in Group 2B (Table IV) and were followed without treatment for a mean of 3.8 years (range, six months to ten years and eight months). These curves were found to have the worst prognosis of any type of congenital scoliosis occurring at any site. All of the scolioses were diagnosed in the first two years of life, when the median curve was 64 degrees (range, 41 to 86 degrees). The curves deteriorated at a median rate of more than 10 degrees per year. All but three of the patients received treatment before the age of five years, when the median curve was 70 degrees (range, 64 to 125 degrees). Two of the three patients who were followed, untreated, for five years received treatment at the age of seven years, with curves of 90 and 148 degrees. The third patient was untreated at ten years old, with a 98-degree curve, which then deteriorated at a rate of 10 degrees per year to become 124 degrees at the age of twelve years and eight months (Figs. 5-A, 5-B, and 5-C).

The most extensive vertebral anomalies in the thoracolumbar region occurred in three patients who had unsegmented bars involving seven to nine vertebrae, with three or four contralateral hemivertebrae. At the age of two years, all of these curves were more than 80 degrees (range, 82 to 103 degrees). One patient was not treated until the age of seven years, when the curve measured 148 degrees.

Pelvic obliquity or listing of the trunk toward the side of the unsegmented bar, or both, occurred in nine of the ten patients with a thoracolumbar curve. The degree of pelvic obliquity was severe (greater than 25 degrees) in two (32 and 50 degrees), moderate (10 to 25 degrees) in four, mild (less than 10 degrees) in two, and absent in two. The listing was severe in four, moderate in one, mild in two, and absent in three.

There were no lumbar curves due to a unilateral un-

segmented bar with contralateral hemivertebrae.

Bilateral Failure of Segmentation (Block Vertebrae) (Group 3)

Block vertebrae were present in thirteen patients. Thus, they were not considered to be a common cause of congenital scoliosis. In our series this type of anomaly most frequently occurred in the upper thoracic region (seven patients) and was least common in the lumbar region (one patient). All of these patients had a single congenital scoliosis (eight left and five right) due to a single block of bilaterally unsegmented vertebrae which ranged from two to five segments in length. Four patients had a block of two vertebrae; six, of three vertebrae; one, of four vertebrae; and two had a block of five vertebrae. No patient required treatment and the mean follow-up was 7.8 years (range, two years to sixteen years and six months), with three patients having reached skeletal maturity. There were nine upper thoracic or lower thoracic curves. All of the curves behaved in the same manner and remained 21 degrees or less (range, 10 to 21 degrees). The rate of deterioration was 1 degree or less per year. Of the three curves that were followed to skeletal maturity, none exceeded 21 degrees. There were three thoracolumbar curves that behaved in a slightly different manner. Two of these curves had virtually no deterioration and measured 10 and 14 degrees when the patients were nine and fourteen years old, respectively. The remaining curve, which was first seen when the patient was ten years old, deteriorated a little and reached 35 degrees at skeletal maturity. The one lumbar curve deteriorated very slowly and measured 24 degrees when the patient was ten years old.

Unilateral Complete Failure of Formation (Hemivertebrae) (Group 4)

Hemivertebrae were the cause of a congenital scoliosis in seventy-seven patients. Two of these patients were seen at skeletal maturity and were in Group 4A. The remaining seventy-five patients were followed without treatment and were in Groups 4B and 4C. The hemivertebrae were always on the convexity of the curve. Seventy patients had a single congenital scoliosis (thirty-six left and thirty-four right), of which fifty-six were due to a single hemivertebra (Table V) and fourteen were due to two hemivertebrae on the same side, producing a single congenital scoliosis (Table VI). Seven patients had hemivertebrae on opposite sides but at different levels of the spine, producing two or more opposing congenital curves (Table II, Cases 4 through 10).

Two patients had a hemivertebra that was initially separate but later became synostosed with one of its neighboring vertebrae. Both were in the lumbar region and the resulting curves progressed less rapidly (less than 1 degree per year) than the majority of the curves in this region. Eight patients had an incarcerated hemivertebra; that is, a small, poorly formed segment of bone tucked into a niche between adjacent normal vertebrae. Four of these in-

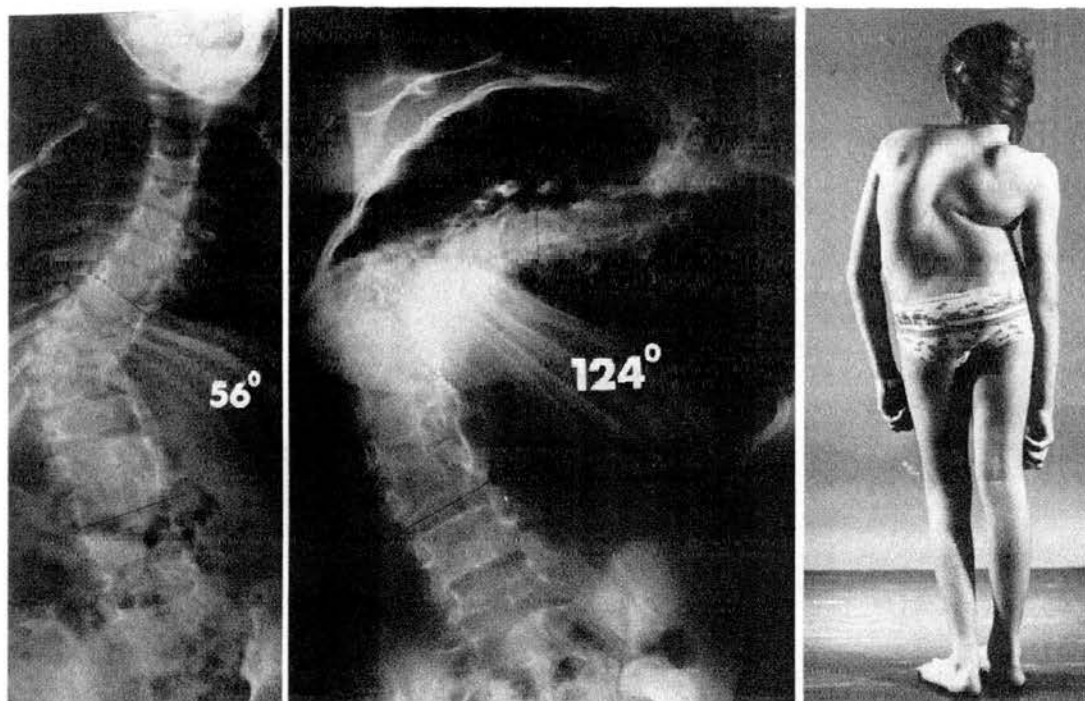


FIG. 5-A

FIG. 5-B

FIG. 5-C

Fig. 5-A: A girl, two years and ten months old, who had a 56-degree left thoracolumbar scoliosis due to a unilateral unsegmented bar on the right extending from the eighth to the twelfth thoracic vertebra with four adjacent contralateral hemivertebrae at the ninth to the twelfth thoracic levels.

Figs. 5-B and 5-C: No treatment was given, and by the age of twelve years and eight months the scoliosis had increased to 124 degrees and the contralateral hemivertebrae were no longer visible. There was pelvic obliquity, and a severe list of the upper part of the body to the right produced a severe cosmetic deformity.

carcerated hemivertebrae occurred in the upper thoracic region; one, in the lower thoracic region; one, in the thoracolumbar region; and two, in the lumbar region. The resulting curves were all less than 20 degrees, with minimum or no progression.

There was a total of 102 hemivertebrae, which were evenly distributed on either side of the spine (fifty-three on the left and forty-nine on the right) and occurred at any level, although they were seen slightly more frequently in the middle thoracic and lower lumbar regions. Of the patients with two unilateral hemivertebrae producing a single congenital curve, the hemivertebrae were separated by four normal vertebrae in two patients, by three normal vertebrae in three patients, by two normal vertebrae in five patients, and by one normal vertebra in three patients; in only one patient were the hemivertebrae adjacent.

Upper Thoracic Curves

Sixteen patients had a single congenital upper thoracic curve, of which thirteen were due to a single hemivertebra (Table V) and three, to two unilateral hemivertebrae (Table VI). Follow-up without treatment was for a mean of four years (range, one year to nine years and ten months). The median rate of deterioration, without treatment, before the age of ten years for the curves due to

a single hemivertebra was 1 degree per year (range, zero to 2 degrees), compared with 2 degrees per year (range, 1 to 2 degrees) in the curves that were due to two unilateral hemivertebrae. By the age of ten years, no curve that was due to a single hemivertebra was greater than 40 degrees, whereas all three patients with two unilateral hemivertebrae had a curve of 40 degrees or more (range, 40 to 47 degrees).

After the patients reached ten years old, the median rate of deterioration doubled (2 degrees per year) in the curves that were due to a single hemivertebra and increased to 2.5 degrees per year (range, 2 to 3 degrees) in those due to two unilateral hemivertebrae. Of six patients with a single hemivertebra, three were followed without treatment to skeletal maturity, at which time the curves measured 25, 39, and 43 degrees, and the remaining three patients had a spine fusion when the curves measured 28, 31, and 44 degrees. Two patients with two unilateral hemivertebrae were followed after the age of ten years and both required a spine fusion during the adolescent growth spurt, when the curves were 43 and 47 degrees.

In general, upper thoracic curves due to one or two unilateral hemivertebrae progressed relatively slowly, and only a few became moderately severe. Although none became very severe, they did, like other congenital upper

TABLE
UNTREATED SINGLE CONGENITAL SCLIOSIS

Group 4B (Patients Seen before the Age of Ten Years)																
Site of Curve	No. of Curves	No.	Age When First Seen* (Yrs.)	Size of Curve (Degrees)*		Rate of Deterioration per Year (Degrees)										
				When First Seen	When Last Seen before 10 Yrs. Old	<1	1	2	3	4	5	6	7	8	9	10
Upper thoracic	13	10	2 (0-7)	16 (11-28)	18 (11-36)	5	3	2								
Lower thoracic	9	6	4 (0-9)	34 (17-48)	44 (17-50)	2		4								
Thoracolumbar	5	3	1 (0-3)	26 (20-35)	40 (21-45)		1	2								
Lumbar	15	12	2 (0-9)	24 (0-37)	30 (13-41)	7	3	2								
Lumbosacral	12	8	4 (2-9)	18 (10-32)	19 (10-53)	6	1		1							

* Median, with range in parentheses.

† The number of patients who were followed untreated until after the age of ten years but who were first seen before the age of ten years, and who are also included in Group 4B, is in parentheses.

thoracic curves, produce a significant cosmetic deformity due to elevation of the shoulder line on the convex side of the curve, and occasionally they produced tilting of the head. In only one patient, with a single hemivertebra at the fifth thoracic level, did a secondary structural lower thoracolumbar scoliosis develop. At skeletal maturity, this curve measured 52 degrees and the upper thoracic congenital curve measured 43 degrees.

Lower Thoracic Curves

Seventeen patients had a single congenital lower thoracic curve, nine of which were due to a single hemivertebra (Table V) and eight, to two unilateral hemivertebrae (Table VI). These patients were followed without treatment for a mean of 3.6 years (range, one year to nine years and four months), and four reached skeletal maturity. The median rate of deterioration before the patients reached the age of ten years was 2 degrees per year (range, zero to 2 degrees) for the curves that were due to a single hemivertebra and 2 degrees per year (range, 2 to 6 degrees) for the curves that were due to two unilateral hemivertebrae. By the age of ten years, the majority of the patients (four of six) with a single hemivertebra had a curve of between 40 and 50 degrees, whereas all of the six patients with two unilateral hemivertebrae had a curve of 50 degrees or more.

After the patients reached the age of ten years, the median rate of deterioration for the lower thoracic curves due to a single hemivertebra increased to 2.5 degrees per year (range, less than 1 degree to 5 degrees), and in the curves due to two unilateral hemivertebrae the median rate was 3 degrees per year (range, 2 to 4 degrees). Of the six patients with a single hemivertebra who were followed after the age of ten years, three reached skeletal maturity without treatment, at which time the curves measured 37, 38, and 43 degrees; two other curves progressed more rapidly and required spine fusion, measuring 52 degrees when one patient was twelve years old and 60 degrees when the other was fourteen years old; and one patient had a 47-degree untreated curve at the age of eleven years. Of the six patients with two unilateral hemivertebrae who

were followed without treatment (Figs. 6-A and 6-B), five required treatment between the ages of twelve and fifteen years, at which time the median curve was 64 degrees (range, 30 to 70 degrees), and one reached skeletal maturity with a 50-degree curve.

Thoracolumbar Curves

Of the ten patients with a thoracolumbar curve, two were in Group 4A. Both of these patients had a single hemivertebra and were seen at skeletal maturity with curves of 52 and 55 degrees. The remaining eight patients with a thoracolumbar curve were followed without treatment for a mean of five years (range, one year and seven months to seven years and seven months).

Five patients had a curve that was due to a single hemivertebra (Table V), and the mean rate of deterioration before the age of ten years was 2 degrees per year (range, 1 to 2 degrees) and 3.5 degrees thereafter (range, 3 to 4 degrees). Before the patients were ten years old, one of three curves was greater than 40 degrees and the two curves that were untreated at skeletal maturity measured 33 and 43 degrees.

Three patients had a single congenital thoracolumbar curve due to two unilateral hemivertebrae (Table VI). These curves were all diagnosed in the first two years of life, when they exceeded 50 degrees. The median rate of deterioration before the age of ten years was 5 degrees per year (range, 4 to 6 degrees) and all three patients required treatment in the third or fourth year of life, when the median curve was 63 degrees.

Lumbar Curves

Fifteen patients had a single congenital lumbar curve, all of which were due to a single hemivertebra at the third or fourth lumbar level (Table V). None of these patients required treatment, and they were followed for a mean of 6.6 years (range, one year to fourteen years and four months). The median rate of deterioration was less than 1 degree per year (range, zero to 2 degrees) before the age of ten years and 1 degree per year thereafter (range, zero to 3 degrees). Before the age of ten years, only one curve was

V

DUE TO A SINGLE HEMIVERTEBRA

Group 4C (Patients Seen at or after the Age of Ten Years)											
No.†	Size of Curve (Degrees)*		Age When Last Seen* (Yrs.)	Rate of Deterioration per Year (Degrees)							
	When First Seen at or after 10 Yrs. Old	When Last Seen or at Maturity		<1	1	2	3	4	5	6	7
6 (3)	30 (15-40)	35 (25-44)	16 (11-17)			4	2				
6 (3)	44 (25-50)	45 (37-60)	16 (11-19)	1	1	1	2		1		
2 (0)	30 (21-40)	41 (33-43)	Both mature				1	1			
8 (5)	32 (14-52)	36 (16-55)	17 (13-18)	2	3	2	1				
12 (8)	20 (15-53)	25 (15-83)	15 (11-18)	5	1	2	3			1	

greater than 40 degrees and six patients had curves that had remained virtually unchanged, at less than 30 degrees, from an early age. Five patients were followed, without treatment, to skeletal maturity, when the median curve was 42 degrees (range, 22 to 55 degrees).

In all of the patients with a lumbar curve the trunk remained balanced, but in two there was mild pelvic obliquity.

Lumbosacral Curves

This type of congenital scoliosis occurred only in association with a single hemivertebra at the lumbosacral junction (Table V). There were twelve such patients, who were followed without treatment for a mean of 6.8 years

(range, one year to ten years and seven months). In nine patients the hemivertebra was at the fifth lumbar level and in three patients the hemivertebra lay between the fifth lumbar level and the sacrum; in one of these patients there was a failure of segmentation between the hemivertebra and the sacrum. The congenital lumbosacral curves were all very short and extended from the fourth or fifth lumbar level to the sacrum. In all of the patients the pelvis remained level, and as a result the hemivertebra caused the lumbar spine to take off obliquely from the sacrum. To overcome this imbalance, in all of the patients a long secondary thoracolumbar curve developed, extending from the fifth lumbar vertebra to the ninth, tenth, or eleventh thoracic vertebra and soon becoming fixed. In eleven pa-

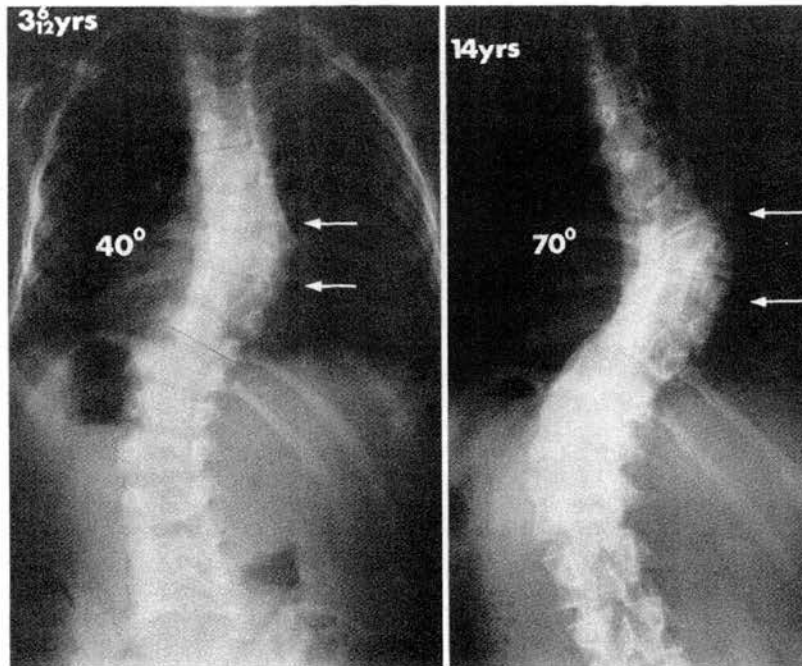


FIG. 6-A

FIG. 6-B

Fig. 6-A: A boy, three years and six months old, with a 40-degree right lower thoracic scoliosis due to two unilateral hemivertebrae at the seventh and tenth thoracic levels.

Fig. 6-B: No treatment was given and the curve deteriorated to 70 degrees at the age of fourteen years, just prior to spine fusion.

TABLE
UNTREATED SINGLE CONGENITAL SCOLIOSIS

Site of Curve	No. of Curves	No.	Age When First Seen* (Yrs.)	Size of Curve (Degrees)*		Rate of Deterioration per Year (Degrees)									
				When First Seen	When Last Seen before 10 Yrs. Old										
						1	2	3	4	5	6	7	8	9	10
Upper thoracic	3	3	6 (3-9)	42 (32-44)	44 (40-47)	1	2								
Lower thoracic	8	4	2 (0-4)	40 (30-48)	53 (50-54)		3				1				
Thoracolumbar	3	3	1 (0-2)	55 (50-60)	63 (55-72)				1	1	1				

* Median, with range in parentheses.

† The number of patients who were followed untreated until after the age of ten years but who were first seen before the age of ten years, and who are also included in Group 4B, is in parentheses.

tients this curve did not compensate sufficiently and as a result the upper part of the body listed to a varying degree to the side opposite that of the hemivertebra. In the one patient in whom the body remained balanced, it did so because of a limb-length discrepancy that resulted in a compensatory pelvic obliquity. With time, all of these secondary curves became structural, and three deteriorated to become the major deformity.

Not all of the secondary curves deteriorated at the same rate, and this appeared to depend on the extent of the hemivertebra. Nine patients had a hemivertebra that extended across the midline, and in these patients both the congenital lumbosacral curve and its secondary thoracolumbar curve remained virtually static until the patients reached the age of ten years, at which time neither curve

exceeded 25 degrees. After the patients were more than ten years old, the rates of deterioration of both curves increased slightly, as did the tendency of the upper part of the body to list to one side. Five of these patients were followed to skeletal maturity, without treatment, at which time neither the congenital lumbosacral curve nor the secondary thoracolumbar curve exceeded 36 degrees and the degree of listing was not sufficient to require treatment.

Three patients had a hemivertebra that did not extend across the midline, and this produced a greater list and a much larger secondary structural thoracolumbar curve, which was severely rotated and produced a large rib hump. Before the patients were ten years old, both the lumbosacral curve and the secondary thoracolumbar curve deteriorated relatively slowly (at a rate of 1 to 3 degrees per

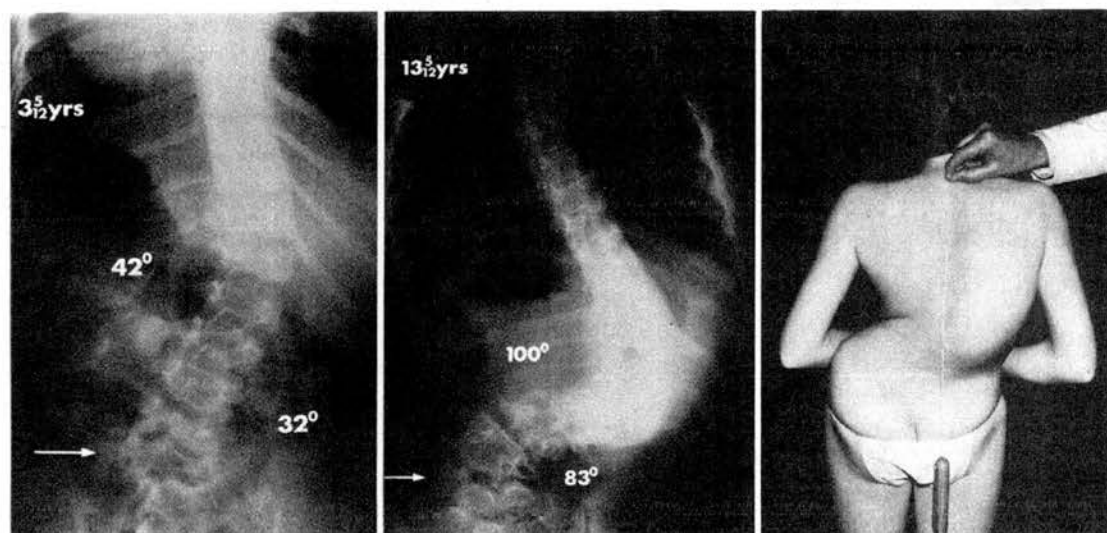


FIG. 7-A

FIG. 7-B

FIG. 7-C

Fig. 7-A: A girl, three years and five months old, with a 32-degree left lumbosacral scoliosis due to a single hemivertebra at the lumbosacral junction. There was also a 42-degree right thoracolumbar scoliosis in which no congenital anomalies were involved. The pelvis was level but the upper part of the body listed to the right.

Figs. 7-B and 7-C: No treatment was given, and by the age of thirteen years and five months the congenital lumbosacral curve had deteriorated to 83 degrees. The thoracolumbar curve measured 100 degrees and had become structural and severely rotated. The list had increased but the pelvis remained level. The major deformity was due to the severe thoracolumbar scoliosis which, despite its size, failed to compensate for the list to the right.

VI

DUE TO TWO UNILATERAL HEMIVERTebrae

Group 4C (Patients Seen at or after the Age of Ten Years)												
No.†	Size of Curve (Degrees)*		Age When Last Seen* (Yrs.)	Rate of Deterioration per Year (Degrees)								
	When First Seen at or after 10 Yrs. Old	When Last Seen or at Maturity		2	3	4	5	6	7	8	9	10
2 (2)	42 (40-44)	45 (43-47)	12 (11-16)	1	1							
6 (2)	51 (28-54)	62 (30-70)	13 (12-17)	2	2	2						
0 (0)	—	—	—									

year), but after the age of ten years a significant cosmetic deformity developed due to the rib hump and an increasing tendency of the upper part of the body to list to one side. The congenital lumbosacral curves in two of these patients measured 25 and 42 degrees at the age of fourteen years, and the secondary structural thoracolumbar curves measured 40 and 48 degrees, respectively. The most severe deformity occurred in the third patient who, at the age of thirteen years, had an 83-degree lumbosacral curve with a 100-degree secondary structural thoracolumbar curve and a severe list (Figs. 7-A, 7-B, and 7-C).

*Unilateral Partial Failure of Formation
(Wedge Vertebrae) (Group 5)*

Wedge vertebrae occurred in only nine patients. They all had a single congenital scoliosis (five left and four right). The most commonly affected vertebrae were in the thoracic region (six patients). Seven patients had a single wedge vertebra and two patients had a pair of adjacent thoracic vertebrae that were affected on the same side.

Upper Thoracic Curves

There was only one upper thoracic congenital scoliosis that was due to wedging, and it involved the fourth thoracic vertebra. This curve deteriorated at a rate of slightly less than 2 degrees per year, from 23 degrees when the patient was twelve years old to 31 degrees at skeletal maturity.

Lower Thoracic Curves

There were four lower thoracic congenital curves, of which two were due to a single wedge vertebra and two, to a pair of adjacent wedge vertebrae. Three patients were followed, without treatment, for a mean of 6.4 years (range, one year and eight months to nine years and nine months) and required treatment at nine, eleven, and twelve years old, when the curves measured 33, 39, and 41 degrees, respectively. One patient with two wedge vertebrae was first seen at skeletal maturity, when the curve measured 49 degrees.

Thoracolumbar Curves

There were three thoracolumbar congenital curves, which were followed for a mean of 3.4 years (range, one year to seven years and four months). One patient required

treatment at the age of five years, when the curve measured 32 degrees. The remaining two patients were untreated, and the curves deteriorated at a rate of 2 degrees per year until they measured 17 degrees, at which time one child was thirteen years old. The other child had a 40-degree curve at skeletal maturity.

Lumbar Curves

Only one lumbar curve was due to wedging, and it involved the fifth lumbar vertebra. This curve deteriorated slowly, at a rate of less than 1 degree per year over three years, and measured 12 degrees when the patient was seven years old.

Complex (Unclassifiable) Anomalies (Group 6)

There were twenty-five patients with complex (unclassifiable) congenital vertebral anomalies causing a congenital scoliosis. Twenty-two of these patients had a single congenital curve (twelve right and ten left) and three had multiple congenital curves.

Nineteen patients had a single congenital scoliosis due to a jumble of vertebral anomalies, and they were followed without treatment for a mean of seven years (range, one to seventeen years). Two patients had an upper thoracic curve; eight patients, a lower thoracic curve; seven patients, a thoracolumbar curve; and two patients, a lumbar curve. All of these curves were much more unpredictable in their behavior than the congenital curves that were due to simple anomalies in the same regions, but in general they tended to progress relatively slowly regardless of their location. Fifteen patients were untreated at the age of ten years, at which time eight curves were less than 20 degrees; three curves, between 20 and 30 degrees; five curves, between 31 and 40 degrees; and only two curves measured more than 40 degrees (51 and 73 degrees). After the age of ten years, the rate of deterioration usually increased. Seven patients reached skeletal maturity without treatment, at which time the curves ranged from 8 to 56 degrees (mean, 31 degrees). The largest untreated curve in the remaining five skeletally immature patients was in the thoracolumbar region and measured 82 degrees at the age of twelve years.

Three patients who were seen at skeletal maturity had a single congenital scoliosis, but the congenital anomaly was partially obscured and could not be accurately

Site of Curvature	Type of Congenital Anomaly					
	Block Vertebra	Wedge Vertebra	Hemivertebra		Unilateral Unsegmented Bar	Unilateral Unsegmented Bar and Contralateral Hemivertebrae
			Single	Double		
Upper thoracic	<1° - 1°	★ - 2°	1° - 2°	2° - 2.5°	2° - 4°	5° - 6°
Lower thoracic	<1° - 1°	2° - 2°	2° - 2.5°	2° - 3°	5° - 6.5°	6° - 7°
Thoraco lumbar	<1° - 1°	1.5° - 2°	2° - 3.5°	5° - ★	6° - 9°	> 10° - ★
Lumbar	<1° - ★	<1° - ★	<1° - 1°	★	> 5° - ★	★
Lumbo sacral	★	★	<1° - 1.5°	★	★	★



No treatment required



May require spinal fusion



Require spinal fusion

★ Too few or no curves

FIG. 8

Median yearly rate of deterioration (in degrees) without treatment for each type of single congenital scoliosis in each region of the spine (187 patients). The numbers on the left in each column refer to patients who were seen before the age of ten years; the numbers on the right refer to patients who were seen at or after the age of ten years.

classified. All three patients had a thoracic or thoracolumbar curve of more than 100 degrees.

Discussion

The prognosis for a patient with congenital scoliosis can vary considerably. Some patients are first seen with small curves, many of which progress minimally, whereas others are first seen with larger curves that deteriorate rapidly and cause extreme deformity. Of the 251 patients in this study, 143 were last seen, untreated, after the age of ten years, at which time fifty-one (36 per cent) had a curve of 40 to 60 degrees and forty (28 per cent) had a curve of more than 60 degrees. We concluded that the prognosis for untreated curves after the age of ten years generally becomes more unfavorable because in our series most of the other 108 patients (that is, an additional sixty-seven patients) required treatment at or before the age of ten years because of the severity of the curve. Twenty of these curves measured 40 to 60 degrees and thirty-nine, more than 60 degrees. These findings are very different from those of Kuhns and Hormell, who found that only 38 per cent of eighty-five children who were followed to skeletal maturity without treatment had curves of more than 30 degrees. Our findings are more in agreement with those of Winter et al., who found that 84 per cent of thirty-eight children who were followed without treatment beyond the age of ten years had curves of more than 40 degrees.

To understand the variable prognosis for congenital scoliosis, it is necessary to correlate the principles of nor-

mal growth of the spine with the pathological anatomy of the various types of congenital vertebral anomalies. Normally, longitudinal growth of the spine is the sum total of the growth occurring at the end-plates on the upper and lower surfaces of the vertebral bodies, which occurs equally on either side of the spine so that the spine remains straight and without a scoliosis². A congenital vertebral anomaly can, however, cause a growth imbalance due to a deficiency in either the number of end plates or their rate of growth on one side of the spine. The lateral curve that results is of a severity proportional to the degree of the growth imbalance.

In classifying a congenital scoliosis with regard to its prognosis, the most important feature is, therefore, the growth imbalance caused by the vertebral anomalies that predominate on one side of the spine. The radiographic classification that was used in this study was found to be generally satisfactory in this respect, although not all of the curves progressed as expected and the growth potential could not be exactly predicted radiographically. The vertebral anomalies in 90 per cent of the patients could be classified into one of five specific groups, but some curves could not be classified (Group 6) because they either involved a complex jumble of anomalies or, on occasion, because the severity was so great as to obscure the radiographic characteristics of the anomaly.

Congenital scoliosis occurred significantly more often in girls than in boys in our series. The congenital curves often were present at birth and were most frequently diag-

nosed in either the first few years of life or between the ages of nine and fourteen years, probably because the periods of most rapid growth of the spine are *in utero*, from birth to the age of three years, and at puberty⁷. The scolioses that presented as a clinical deformity in the first year of life had the worst prognosis, as this indicated a marked growth imbalance that would continue until skeletal maturity, resulting in severe deformity.

A single congenital scoliotic curve occurred in 95 per cent of the 251 patients and was much more common than the occurrence of two or more congenital curves (5 per cent of the patients). The commonest type of vertebral anomaly causing the scoliosis was a unilateral unsegmented bar (38 per cent of the 269 congenital curves; Table I), followed by hemivertebrae (33 per cent), complex anomalies (11 per cent), a unilateral unsegmented bar with contralateral hemivertebrae (10 per cent), and a block vertebra (5 per cent); the least common was a wedge vertebra (3 per cent). The commonest site for the congenital scoliosis was the lower thoracic region (33 per cent of the 269 congenital curves; Table I), followed by the upper thoracic (31 per cent), thoracolumbar (20 per cent), and lumbar regions (11 per cent); the least common was the lumbosacral region (5 per cent).

Like Winter et al., we found that the rate of deterioration and the ultimate severity of the congenital scoliosis depended not only on the type of anomaly but also on the site at which it occurred. The site of curvature that had the worst prognosis, for each type of vertebral anomaly, was usually the thoracolumbar region, and the prognosis was only slightly less severe in the lower thoracic region. These findings differ from those of Winter et al., who found that lower thoracic curves had a worse prognosis than thoracolumbar curves. We agree with Winter et al. that lower thoracic and thoracolumbar curves usually have a worse prognosis than do lumbar curves, and that the most benign curves occur in the upper thoracic region.

The type of anomaly causing the most severe scoliosis in each region of the spine was a unilateral unsegmented bar with contralateral hemivertebrae at the same level. This was followed in severity by scoliosis caused by a unilateral unsegmented bar alone, two unilateral hemivertebrae, a single hemivertebra, and a wedge vertebra; the least severe was scoliosis caused by a block vertebra (Tables II through VI).

In addition, we found that the rate of deterioration of the curves was not constant, but if the curve was present before the patient was ten years old it usually increased, in particular during the adolescent growth spurt. Although there was often a relatively wide range in the rate of deterioration, the majority of the curves that were due to radiographically similar anomalies and that occurred in the same region tended to deteriorate at approximately the same rate (Fig. 8).

A unilateral unsegmented bar does not contain growth plates and therefore cannot grow longitudinally, whereas normal or nearly normal growth may occur on the opposite

side of the spine. In our series, the longer unsegmented bars tended to produce the larger curves in a specific region, but occasionally a shorter bar produced an equally large curve in the same region. In three children the unsegmented bar was not recognized radiographically until they were three and four years old, when it became more ossified. As a result, these patients were initially misdiagnosed as having infantile idiopathic scoliosis. Because of the severe growth imbalance, all of the curves that were associated with an unsegmented bar and that were present during infancy deteriorated very rapidly, and all became very severe. The mean rate of deterioration in patients who were younger than ten years old ranged from 5 degrees per year for lower thoracic curves to 6 degrees per year for thoracolumbar curves (Table III). Of the thirty-six patients with a lower thoracic, thoracolumbar, or lumbar curve who were last seen without treatment at or before the age of ten years, ten had a curve that measured between 40 and 60 degrees and nineteen had a curve of more than 60 degrees. This type of congenital scoliosis requires treatment as soon as the anomaly is diagnosed.

It is important to recognize the anomaly of a unilateral unsegmented bar with contralateral hemivertebrae, as described by Nasca et al., because it has the worst prognosis of any type of congenital vertebral anomaly. All of these patients were seen before the age of two years in our series. At that age the hemivertebrae could be seen radiographically, but as the deformity progressed the hemivertebrae tended to be obscured because of the severity of the curve, and the anomaly then became indistinguishable from an unsegmented bar alone. The median rate of deterioration before the age of ten years ranged from 5 degrees per year for upper thoracic curves to more than 10 degrees per year for thoracolumbar curves. Sixteen of eighteen lower thoracic and thoracolumbar curves exceeded 50 degrees once the patient was three years old (Table IV). This type of congenital scoliosis requires treatment as soon as the anomaly is diagnosed.

A hemivertebra produces a scoliosis by acting as an enlarging wedge on the affected side of the spine, whereas in patients with a unilateral unsegmented bar there is retarded growth on the affected side. The growth imbalance in patients with hemivertebrae is never as severe as in those with a unilateral unsegmented bar. Not all hemivertebrae, however, produce the same degree of growth imbalance at the same site. Eight patients in our series had an incarcerated hemivertebra⁸, which we defined as a small, poorly formed, extra segment of bone tucked into the spine between adjacent normal vertebrae. This type of hemivertebra has no growth potential and the resulting curves were all less than 20 degrees, with minimum or no progression. Two patients had a hemivertebra that became synostosed with a neighboring vertebra (semisegmented), and this also produced a curve that progressed less rapidly than the others in the same region. The majority of single hemivertebrae, however, caused slowly progressive curves. Before the patients were ten years old the median

rate of deterioration for both upper thoracic and lumbar curves due to a single hemivertebra was 1 degree per year and no curve exceeded 40 degrees (Table V). These curves usually do not require treatment. Single hemivertebrae in the lower thoracic and thoracolumbar regions caused more severe deformity. Before the patients were ten years old, the median rate of deterioration was 2 degrees per year, and six of nine curves were between 40 and 50 degrees. These patients often require treatment during the adolescent growth spurt. Two unilateral hemivertebrae produced a greater growth imbalance and caused a much more severe deformity. The median rate of deterioration before the age of ten years ranged from 2 degrees per year for upper thoracic curves to 5 degrees per year for thoracolumbar curves. All seven lower thoracic and thoracolumbar curves were greater than 50 degrees by the time the patient was ten years old. These curves usually require treatment before the patient is five years old.

A wedge-shaped vertebra is due to a unilateral partial failure of formation of a vertebra, with retarded longitudinal growth on the hypoplastic side. In two patients, two adjacent vertebrae were affected, but this did not unduly increase the severity of the scoliosis. The one upper thoracic curve and one lumbar curve in our series deteriorated slowly (less than 2 degrees per year) and did not require treatment, whereas the seven lower thoracic and thoracolumbar curves deteriorated somewhat more rapidly (median rate of increase, 2 degrees per year) and three required treatment during the adolescent growth spurt.

A block vertebra is due to bilateral failure of segmentation, and longitudinal growth is impaired on both sides of the spine but not always symmetrically. The result was a mild degree of curvature that rarely exceeded 20 degrees, and the thirteen patients in our series who were so affected did not require treatment.

The thirteen patients who had two or more congenital curves had a variable prognosis, depending on the site of the opposing anomalies. If the anomalies, either similar or dissimilar, occurred within a few segments of each other and were in the same region, they tended to balance each other and produced little deformity other than a kink in the spine. If, however, they were widely separated and in different regions, the resulting curves tended to be unbalanced, producing decompensation or listing of the trunk that required treatment.

Apart from the rate of deterioration and ultimate severity of a congenital scoliosis, there were also a number of important secondary features that should be emphasized because they contributed significantly to the over-all disability and deformity of the patient.

Upper thoracic curves, especially those that extended cranially beyond the cervicothoracic junction, commonly produced a cosmetic deformity due to elevation of the shoulder or, less frequently, tilting of the head (Figs. 2-A and 2-B). Because congenital scoliosis occurs so frequently in the upper thoracic region (31 per cent of our patients) the deformity is a common one, and the higher the

apex of the curve the more severe was the deformity. An elevated shoulder line was most distressing to girls, and a 30-degree curve seemed to be the upper limit that the patients would tolerate.

An additional problem associated with thoracic curves, especially those with the apex at the fifth, sixth, or seventh thoracic vertebra, was the development of a long secondary structural curve in the lower thoracic or thoracolumbar region. This curve, which did not involve any congenital anomalies, initially was compensatory and was correctable, but later it tended to become fixed and to deteriorate even more rapidly than the primary (congenital) curve (Figs. 3-A and 3-B). Moreover, the congenital thoracic curve usually had only a mild degree of rotation, whereas the lower secondary curve often was severely rotated. The large rib hump so produced usually constituted a major deformity. In nineteen of forty-five patients who had a congenital curve due to a unilateral unsegmented bar, with or without contralateral hemivertebrae in the upper thoracic segments, a secondary structural lower thoracic or thoracolumbar curve developed.

In patients with a lower thoracic, thoracolumbar, or lumbar curve, especially those due to a unilateral unsegmented bar with or without contralateral hemivertebrae, a compensatory curve that was adequate to balance the congenital curve often failed to develop because there were too few normal mobile vertebrae between the anomaly and either the upper end of the spine or the sacrum. As a result, in 80 per cent of the patients with a congenital thoracolumbar curve and in all with a lumbar curve in our series some degree of pelvic obliquity and an apparent shortening of one lower limb developed (Figs. 5-B and 5-C). Decompensation or listing of the upper part of the body to one side was also a common finding associated with the more severe lower thoracic and thoracolumbar curves and could result in a very severe deformity.

The twelve lumbosacral curves in our series were all due to a single hemivertebra at the lumbosacral junction but, unlike the situation with congenital lumbar or thoracolumbar curves, the pelvis remained level. The hemivertebra, therefore, caused the lumbar spine to take off obliquely from the sacrum. In an attempt to keep the body balanced, a secondary lumbar or thoracolumbar curve developed, but unfortunately it usually was insufficient to prevent the trunk from listing to the side opposite that of the hemivertebra. In nine patients, both the list and the secondary curve remained small (less than 36 degrees) and produced only a mild to moderate deformity. In three patients, however, the secondary curve became large, fixed, and rotated and caused a major cosmetic deformity due to a large rib hump and decompensation of the trunk (Figs. 7-B and 7-C).

In conclusion, we can state that congenital scoliosis is a potentially serious condition, which can and often does result in severe curvature of the spine and malalignment of the body. We have shown that it is possible to anticipate the course of a congenital scoliosis if the type of vertebral

anomaly and its site are known. Ideally a congenital scoliosis should be diagnosed while the patient is young and the curve is small. At that stage, a curve that is at risk for progression can be recognized and an appropriate prophylactic course of treatment can be planned so as to prevent severe deformity.

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Occult Intraspinous Anomalies and Congenital Scoliosis

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ABSTRACT: Of 251 patients with congenital scoliosis, occult congenital intraspinal anomalies were diagnosed in forty-six (18.3 per cent). A diastematomyelia was the commonest anomaly (forty-one patients). Other less common anomalies, occurring alone or in association with a diastematomyelia, were: neurenteric, epidermoid, and dermoid cysts; teratoma; lipofibroma; absence of nerve roots; fibrous bands; and a tight filum terminale. Intraspinous anomalies were associated with all types and sites of congenital scoliosis but by far the highest incidence (52 per cent) occurred in association with a unilateral unsegmented bar with contralateral hemivertebrae in the lower thoracic or thoracolumbar regions. Thirty of the patients with an intraspinal anomaly had neural abnormalities, which usually affected only one lower extremity, and in twenty-four patients a paralytic foot deformity developed. Neural deterioration occurred in nine of these patients before the age of five years and was halted by excision of the anomaly. An additional twelve patients (4.8 per cent) of the 251 with congenital scoliosis also had a unilateral neural deficit in the lower limb and a paralytic foot deformity, similar to those found in the patients with an intraspinal anomaly, but had no myelographic evidence of a structural anomaly.

The purpose of this paper is to report the incidence and describe the types of occult intraspinal anomalies and neural abnormalities in the lower limb in patients with congenital scoliosis.

The development of the spinal cord is closely associated with that of the vertebral column, and it is not surprising that neural and vertebral malformations often coexist. Vertebral anomalies can produce an imbalance in the longitudinal growth of the spine and result in a progressive congenital scoliosis^{7,10}. Because of the relationship between neural and vertebral development, one would therefore expect to find a high incidence of intraspinal abnormalities in association with congenital scoliosis. These intraspinal anomalies may not be recognized, however, because the skin overlying the spine usually appears normal and the associated neural abnormalities are mild or absent. The various types of occult congenital intraspinal anomalies in patients without scoliosis have been well described by James

and Lassman. The most common by far is a diastematomyelia, which is defined as a partial or complete sagittal split in a localized segment of either the spinal cord or the cauda equina. Lying within the split there always is an osseous or fibrocartilaginous spur that projects backward in the midline from the posterior aspect of one or more adjacent vertebral bodies. The spur invaginates the dura, and while the neural elements of the cord always are joined together just above the spur, they may or may not be joined below it. Other congenital intraspinal anomalies, less common than diastematomyelia, are epidermoid cysts, dermoid cysts, neurenteric cysts, lipomas, and teratomas⁵. The spinal cord or cauda equina can also be tethered by fibrous bands, a tight filum terminale, ectopic posterior nerve roots, or arachnoid adhesions to the dura or vertebral column.

In patients with scoliosis and occult intraspinal anomalies, the normal movement of the spinal cord within the spinal canal may be restricted, and any attempt to correct the scoliosis could result in stretching of the spinal cord and serious neural complications⁹. Therefore, before correcting a scoliosis it is essential to detect any intraspinal anomaly.

Materials and Methods

I studied the cases of 251 patients with congenital scoliosis who attended the Edinburgh Scoliosis Clinic between 1958 and 1981. Patients with a myelomeningocele were excluded. The majority of patients were seen after 1965, and their ages at the time of diagnosis of the scoliosis ranged from birth to twenty-two years. McMaster and Ohtsuka have reported on the natural history of the spinal deformity in these 251 patients.

The spine radiographs of the 251 patients were examined for the presence of a midline bone spur indicative of a diastematomyelia or for any other manifestation of an intraspinal anomaly. Myelograms were available for 106 of the 251 patients. Myelography was performed on all patients with clinical or radiographic evidence that made us suspicious of an intraspinal anomaly, and in the last twelve years it has been done in all patients who were about to undergo surgery to correct the scoliosis.

The case records of all 251 patients were reviewed for the presence of structural or neural abnormalities affecting the lower limbs, bladder, or bowel, and photographs were reviewed for cutaneous abnormalities overlying the spine.

Fifty-eight patients from the total of 251 are the subject of this report. They fell into two groups: forty-six patients who were found to have a structural intraspinal anomaly

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and twelve patients with abnormal neural findings but no myelographic evidence of an intraspinal anomaly.

Table I shows the details of fourteen patients whose cases were of special interest. Nine of them (Cases 1 through 9) had other types of intraspinal anomalies in addition to or without a diastematomyelia, and five patients (Cases 10 through 14) had a progressive neural abnormality affecting the lower limbs or the bladder.

Intraspinal Anomalies

Of the forty-six patients with a congenital intraspinal anomaly, thirty-nine had a myelographic examination. In the other seven, the routine radiographs showed a bone spur and the diagnosis of a diastematomyelia was not in doubt. The diagnosis was confirmed by spinal exploration in thirty-five patients. Of the eleven patients who did not have the intraspinal anomaly surgically explored, ten had a diastematomyelia with a stable neural deficit and were seen before 1970, when the possible complications due to the spur were not fully appreciated. The eleventh patient (Case 7), who was seen more recently, had a unilateral absence of nerve roots that did not require surgical treatment.

Of the thirty-five patients who had the intraspinal anomaly surgically treated, thirty-one had a diastematomyelia: a bone spur was removed from twenty-seven and a fibrous spur, from four. In addition to the diastematomyelia, four patients (Cases 1 through 4) had other types of intraspinal anomalies, which included a neurenteric cyst, an epidermoid cyst, a dermoid cyst, and fibrous adhesions of the cauda equina. The remaining five patients (Cases 5 through 9) did not have a diastematomyelia but did have other types of intraspinal anomalies, which included a neurenteric cyst, lipofibromas, a complex teratoma, and fibrous adhesions of the cauda equina in conjunction with a tight filum terminale.

Clinical Presentation

At the time when the intraspinal anomalies were diagnosed (Fig. 1), most of the patients (twenty-four of forty-six) were less than seven years old. Forty of the patients were girls, two of whom were sisters. There were six boys. Although all of the patients had scoliosis, and most of them were seen because of that deformity, there were other reasons why treatment was sought.

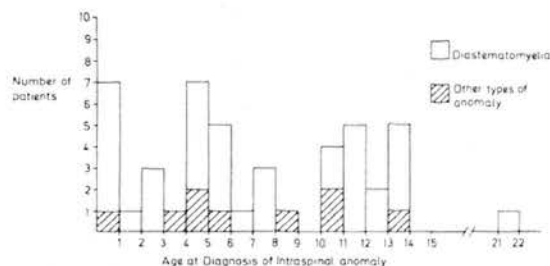


FIG. 1

Age at the time of diagnosis of the intraspinal anomaly.

Six patients were seen because of an abnormality of the skin or subcutaneous tissues overlying the spine. A hairy patch was present in four patients and a subcutaneous lipoma, in two. When first seen, all but one of these patients were infants (mean age, one year). Two patients were also found to have a unilateral neural abnormality, and a foot deformity developed in another over the next few years. Scoliosis was present in four patients when they were first seen, but it did not develop in the remaining two patients until later in childhood. All were found to have a diastematomyelia and one (Case 1) also had a neurenteric cyst at a higher level.

Eight patients had a deformity of the foot that developed because of the neural deficit and the muscle imbalance secondary to it. The deformity was unilateral in six patients and bilateral in two. When first seen, all of these eight patients were less than five years old (mean age, three years). The youngest was one month old. The neural abnormality remained static in four patients who were followed for a mean of five years and five months (range, one to nine years) without treatment. Three of these patients had a diastematomyelia and one (Case 8) had fibrous adhesions of the cauda equina and a tight filum terminale. In the remaining four patients (Cases 5, 6, 9, and 13) the neural abnormalities continued to deteriorate, resulting in increasing muscle weakness and foot deformity. Urinary incontinence developed in two patients (Cases 5 and 9) in the second and third years of life. The intraspinal anomaly was excised from all four of these patients between the ages of three years and six months and eight years and nine months (mean age, six years and one month).

In one patient (Case 14) urinary incontinence had developed at the age of four years. She had scoliosis and a unilateral neural abnormality affecting the lower limb with mild unilateral clawing of the toes. A bone spur associated with a diastematomyelia was excised immediately.

The remaining thirty-one patients were first seen because of the scoliosis. Fifteen of these patients also had a neural abnormality at initial examination. The neural deficit was unilateral in thirteen patients and bilateral in two. Deformity and weakness of the foot was present in nine patients, and was bilateral in two. None of these fifteen patients had urinary incontinence. A diastematomyelic spur was immediately excised from nine patients, as well as an epidermoid cyst at the same level in one (Case 2). This was done as a prophylactic measure to prevent the possible development of further neural abnormalities during correction of the scoliosis.

Six patients who were seen at a time when the possible problems associated with an intraspinal anomaly were not fully appreciated did not have any treatment. They were followed for a mean of four years (range, one year to six years and six months) and their neural status remained unchanged. Five had a diastematomyelia and one (Case 7) had unilateral absence of nerve roots.

Seven patients with scoliosis were normal neurologically, but a bone spur indicative of a diastematomyelia was

TABLE
DETAILS ON FOURTEEN PATIENTS

Case	Mode of Presentation	Sex, Age (Yrs. + Mos.)	Intraspinal Anomaly: Type and Site	Cutaneous Abnormality over Spine	Lower- Extremity Deformity	Lower Extremity Neural Abnormalities			
						Motor	Sensory	Reflex*	Bladder
1	Subcutaneous lipoma over spine	F, 0 + 8	Neurenteric cyst T3 to T4, bone spur L1 to L2	Lipoma; thoracolumbar	None	None	None	None	None
2	Scoliosis	F, 5 + 3	Epidermoid cyst and bone spur L4 to L5	Hairy patch	None	None	None	R AJ and KJ	None
3	Scoliosis	M, 1 + 2	Dermoid cyst and bone spur L4 to L5	Hairy patch; lumbar	Equinovarus, R foot; hypoplasia, R lower limb	R lower limb	None	Bilat. AJ, KJ, and Babinski	None
4	Scoliosis	F, 2 + 10	Fibrous spur T12 to L2, fibrous adhesions of cauda equina	None	Hypoplasia, R lower limb	None	None	None	None
5	Foot deformity	F, 3 + 4	Neurenteric cyst T4 to T6, lipofibromas T11 to L2	Hairy patch; upper thoracic and lumbar	Bilat. equinovarus	Bilat. lower limbs	None	Bilat. AJ, KJ, and Babinski	Incontinent
6	Foot deformity	F, 4 + 10	Complex teratoma T9 to L4	Lipoma; thoracolumbar	Pes cavus and claw toes, L foot; hypoplasia, L lower limb	L lower limb	L foot	L AJ and KJ, bilat. Babinski	None
7	Scoliosis	M, 10 + 0	Unilat. absence of nerve roots L5 and S1 on right	None	Pes cavus and claw toes, R foot; hypoplasia, R lower limb	R lower limb	R foot	R AJ and KJ	None
8	Foot deformity	F, 4 + 1	Fibrous adhesions of cauda equina and tight filum terminale	Hairy patch; thoracolumbar	Equinovarus, L foot; hypoplasia, L lower limb	L lower limb	L foot	L AJ and Babinski	None
9	Foot deformity	F, 0 + 9	Fibrous adhesions of cauda equina and tight filum terminale	Hairy patch, pigmented nevus, dimple; lumbar	Equinovarus, L foot; hypoplasia, L lower limb	L lower limb	L foot	L AJ, KJ, and Babinski	Incontinent
10	Scoliosis	F, 2 + 1	Bone spur T12	None	Pes cavus, L foot; hypoplasia, L lower limb	L foot	None	L Babinski	Incontinent
11	Scoliosis	F, 0 + 2	Bone spur L2	Hairy patch; mid-thoracic	Pes cavus, R foot; hypoplasia, R lower limb	R lower limb	R foot	R AJ, KJ, and Babinski	None
12	Scoliosis	F, 3 + 10	Bone spur T10	None	Pes cavus, L foot; hypoplasia, L lower limb	L lower limb	None	L AJ and Babinski	Occas. incontinent
13	Foot deformity	F, 1 + 8	Bone spur L1	Hairy patch; lumbar	Pes cavus, L foot; hypoplasia, L lower limb	L lower limb	None	L AJ and KJ	None
14	Urinary incontinence	F, 4 + 0	Bone spur T8 to T10	Hairy patch; lumbar	Claw toes, L foot	L foot	None	L AJ, KJ, and Babinski	Incontinent

* AJ = ankle jerk and KJ = knee jerk.

clearly seen on the plain radiographs of the spine that were made at their initial assessment. The spur was immediately excised as a prophylactic measure from three of these patients before the age of five years (mean age, one year and nine months). Four patients had no treatment for the intraspinal anomaly and were followed to skeletal maturity

without development of a neural abnormality.

A further five patients with scoliosis also were neurologically normal, and they were followed for a mean of six years and eight months (range, one year to ten years and six months) without development of a neural abnormality. A diastematomyelia was diagnosed only when a

I

WITH AN OCCULT INTRASPINAL ANOMALY

Vertebral Anomaly Type	Scoliosis		Treatment, Age (Yrs. + Mos.)		Size of Curve (Degrees)		Neural Change Since First Seen	Age at Last Follow-up (Yrs.)
	Site	Site and Type of Curve	Intraspinal Anomaly	Scoliosis	Before Fusion or When Last Seen	After Fusion		
Unilat. unseg. bar and contralat. hemivertebrae	T11-L2	R thoracolumbar	Excised, 1 + 0	Milwaukee brace, 2 + 1	64		No change	8
Unilat. unseg. bar and contralat. hemivertebrae	T1-T10	L lower thoracic	Excised, 5 + 7	Post. fusion in Risser jacket, 6 + 1	72	72	No change	8
Unilat. unseg. bar and contralat. hemivertebrae	T8-L1	L thoracolumbar	Excised, 4 + 9	Post. fusion with Harrington instrumentation, 9 + 2	90	71	Early neural deterioration; no change after excision of anomaly	12
Unilat. unseg. bar and contralat. hemivertebrae	T9-L1	L thoracolumbar	Excised, 13 + 1	Ant. vert. resection, 13 + 5; post. fusion with Harrington instrumentation, 13 + 6	124	80	Partial motor paralysis of R lower limb following ant. vert. resection	14
Unilat. unseg. bar	T12-L3	L lumbar	Excised, 3 + 6	Post. fusion with Harrington instrumentation, 12 + 5	57	45	Early neural deterioration; incontinence improved after excision of anomalies	15
Complex anomalies	T4-L2	R lower thoracic	Excised, 8 + 9	No treatment	50		Early neural deterioration; partial sensory recovery after excision of anomalies	17
Hemivertebra	L5	R lumbosacral	No treatment	No treatment	17		No change	13
Unilat. unseg. bar and contralat. hemivertebrae	T3-T9	R lower thoracic	Excised, 10 + 0	Post. fusion with Harrington instrumentation, 10 + 1	72	45	No change	12
Unilat. unseg. bar and contralat. hemivertebrae	T11-L2	R thoracolumbar	Excised, 4 + 11	Ant. vert. resection, post. fusion with Harrington instrumentation, 6 + 5	148	90	Early neural deterioration; no change after ex- cision of anomalies	10
Unilat. unseg. bar and contralat. hemivertebrae	T5-T10	R lower thoracic	Excised, 5 + 10	Post. fusion with Harrington instrumentation, 10 + 4	71	45	Early neural deterioration; incontinence resolved after excision of spur	16
Complex anomalies	T4-L3	R lower thoracic	Excised, 6 + 0	Post. fusion in Risser jacket, 10 + 6	39	37	Early neural deterioration; no change after excision of spur	14
Hemivertebrae	T10, T12	R thoracolumbar	Excised, 5 + 2	Post. fusion in Risser jacket, 10 + 9	40	32	Early neural deterioration; incontinence resolved after excision of spur	18
Complex anomalies	T4-T7	L thoracolumbar	Excised, 7 + 1	Post. fusion with Harrington instrumentation, 11 + 9	94	64	Early neural deterioration; no change after excision of spur	15
Unilat. unseg. bar	T9-L1	L thoracolumbar	Excised, 4 + 0	Post. fusion with Harrington instrumentation, 15 + 8	53	32	Urinary incontinence resolved after excision of spur	18

myelogram was made as a routine investigation prior to correction and fusion of the scoliosis, between the ages of ten and fourteen years.

The remaining four patients (Cases 3, 10, 11, and 12) who were seen because of scoliosis had the intraspinal anomaly diagnosed when a progressive neural abnormality developed during follow up. When first seen, three of these

patients were infants with apparently normal neurological findings. Over the next few years progressive unilateral neural abnormalities, muscle weakness, and ipsilateral foot deformities developed. Urinary incontinence also developed in one patient at the age of five years. The fourth patient was first seen at the age of three years and ten months with a very mild unilateral neural abnormality of the lower limb.

TABLE II
SITES AND TYPES OF CONGENITAL SCLIOSIS IN TWO HUNDRED AND FIFTY-ONE PATIENTS
AND THE NUMBER WITH AN OCCULT INTRASPINAL ANOMALY

Site of Curve	Type of Congenital Vertebral Anomaly* (No. of Patients)					
	Unilateral Unsegmented Bar	Unilateral Unsegmented Bar with Contralateral Hemivertebra	Block Vertebra	Hemivertebra	Wedge Vertebra	Unclassifiable Complex Anomalies
Upper thoracic	38 (3)	7 (2)	7 (0)	16 (1)	1 (0)	2 (0)
Lower thoracic	38 (6)	11 (5)	2 (1)	17 (0)	4 (1)	10 (1)
Thoracolumbar	15 (7)	10 (6)	3 (0)	10 (1)	3 (0)	9 (3)
Lumbar	5 (1)	0	1 (1)	15 (0)	1 (0)	2 (1)
Lumbosacral	0	0	0	12 (4)	0	0
Multiple curves	3 (1)	0	0	7 (0)	0	3 (1)
Total	99 (18)	28 (13)	13 (2)	77 (6)	9 (1)	25 (6)

* Numbers in parentheses indicate the numbers with an occult intraspinal anomaly.

A mild foot deformity developed and she became incontinent of urine at the age of five years. The intraspinal anomaly was excised from all four patients between the ages of five and seven years. All were found to have a diastematomyelia and one child (Case 3) also had a dermoid cyst at the same level.

Cutaneous Abnormalities

Thirty-four (74 per cent) of the forty-six patients with intraspinal anomalies had one or more abnormalities of the skin or subcutaneous tissues overlying the spine. These abnormalities usually occurred in the same region as the intraspinal anomaly, but in four patients they occurred at a different site. The most common cutaneous abnormality was a hairy patch (twenty eight patients), which ranged from a poorly demarcated, very light, downy hirsuteness to a large, often triangular patch, with long thick hair. The hairy patch was associated with a pigmented nevus in three patients, with a dermal sinus in one patient, and with a combined pigmented nevus and dermal sinus in one patient. A pigmented nevus occurred by itself in two patients and in association with a dermal sinus in one patient. A subcutaneous lipoma occurred in three patients (Figs. 4-A and 5-A).

Neural Abnormalities

When first seen, twenty-seven of the forty-six patients with intraspinal anomalies were found to have a neural deficit in one or both lower limbs or in the bladder. An additional three patients, first seen shortly after birth, were thought to be neurologically normal, but a neural deficit became apparent within a year.

The neural deficit was progressive in nine patients, all of whom were seen before the age of five years. Five of these patients (Cases 10 through 14) had a diastematomyelia and one (Case 3) had a diastematomyelia and a dermoid cyst. The remaining three patients with a progressive deficit had other types of intraspinal anomalies: a neurenteric cyst and lipofibromas (Case 5), a teratoma (Case 6), and fibrous adhesions of the cauda equina (Case 9).

The neural deficit did not progress in ten patients who

were first seen at a mean age of six years and nine months (range, two to thirteen years) and who were followed without treatment for a mean of six years and two months (range, ten months to fifteen years). The remaining eleven patients with a deficit had the intraspinal lesion treated surgically as prophylaxis soon after they were first seen (mean age, eight years and three months; range, one month to twenty-one years).

The neural deficit affected one lower limb in twenty-two patients (nine right and thirteen left) and both lower limbs in eight patients although the deficit usually was much more pronounced in one limb than in the other. Of the eight patients in whom both lower extremities were affected, five had a diastematomyelia; one, a diastematomyelia and a dermoid cyst (Case 3); one, a neurenteric cyst and lipofibromas (Case 5); and one, a complex teratoma (Case 6). The most common deficits in reflexes were an absent or diminished ankle jerk (twenty-eight patients), an abnormal knee jerk (twenty patients), and a positive Babinski sign (twenty-one patients).

Sensory deficits, varying in degree and affecting mainly the fore part of the foot, were present unilaterally in fourteen patients, but sensory testing in young children is difficult and it is possible that some additional patients may have had a mild deficit.

Urinary incontinence was present in six patients before the age of five years and it did not develop in any patient after the age of five.

Muscle weakness was present in one lower limb in twenty two patients and in both lower limbs in three. Rarely was there a complete paralysis of any muscle group, and usually only the muscles distal to the knee were affected, most commonly the evertors and dorsiflexors of the foot.

Deformities

A mild unilateral generalized underdevelopment and shortening of the lower limb and foot occurred on the same side as the abnormal neurological findings in twenty four patients. There were also eight patients in whom one lower limb was similarly affected who had no neural deficit.

A deformity of the foot occurred in twenty-four patients and was unilateral in all but three. Ten patients had pes cavus with clawing of the toes and an inverted heel, seven had an equinovarus deformity, five had clawing of the toes alone, and two had a calcaneovalgus deformity.

The data on scoliosis in the pool of 251 patients are shown in Table II, as compared with the patients who had an intraspinal anomaly. Four patients with an intraspinal anomaly did not have scoliosis when they were first seen. These four patients, all less than four years old, were seen either with a cutaneous abnormality overlying the spine or with a deformity of the foot, and a scoliosis developed in all of them by the age of nine years. In many other patients with an intraspinal anomaly scoliosis never develops, and therefore they are not included in this report.

In the total pool of 251 patients with congenital scoliosis, an intraspinal anomaly was most commonly found in the twenty-eight patients with a curve due to a unilateral unsegmented bar with a contralateral hemivertebra at the same level. Thirteen (46 per cent) of these patients had an intraspinal anomaly. The other types of congenital scoliosis were associated with an intraspinal anomaly less often; the association was present in six (24 per cent) of the twenty-five patients with an unclassifiable complex anomaly, in eighteen (18 per cent) of the ninety-nine patients with a unilateral unsegmented bar, and in six (8 per cent) of the seventy-seven patients with a hemivertebra. Two of the thirteen patients with a block vertebra and one of the nine with a wedge vertebra had an associated intraspinal anomaly.

Of the forty-six patients with an intraspinal anomaly, the commonest region of the spine affected by the congenital scoliosis was either thoracolumbar (seventeen patients; 37 per cent) or lower thoracic (fourteen patients; 30 per cent). The commonest type of vertebral anomaly causing the scoliosis in these forty-six patients was a unilateral unsegmented bar (eighteen patients; 39 per cent) or a unilateral unsegmented bar with a contralateral hemivertebra at the same level (thirteen patients; 28 per cent).

The rate of deterioration of the congenital curves in the patients with an intraspinal anomaly did not differ significantly from the rate of similar curves in patients without an intraspinal anomaly⁷. Surgical treatment of the intraspinal anomaly did not prevent or inhibit deterioration of the scoliosis in fourteen patients prior to spine fusion or in the five patients who did not have a fusion.

Radiographic Features of the Intraspinal Anomalies

In thirty-three of the forty-one patients with diastematomyelia a bone spur could be identified. It usually consisted of a small oval density (Fig. 3-A) lying in the midline. However, as the scoliosis deteriorated and became more distorted, the spur became increasingly more difficult to distinguish, and in some patients it was only by means of myelography that we could visualize the spur. In twenty-one patients the spur was at the same spinal level as the abnormalities in the vertebral bodies that produced the con-

genital scoliosis. In seventeen of the remaining twenty patients the spur was at a lower level while in three patients, all of whom had a lumbosacral hemivertebra, the spur was at a higher level. The number of normal vertebrae between the spur and the vertebral anomalies producing the scoliosis was one or two in nine patients, four or five in four patients, six or seven in three patients, and eight to twelve in four patients.

Spurs occurred at any level from the fourth thoracic to the fifth lumbar vertebra, but were most common in the lumbar region (Fig. 2). The spur projected from one vertebra in twenty-nine patients, from two in nine patients, and from three in three patients. No patient had two spurs. The most commonly affected level was the first lumbar (sixteen patients; 39 per cent).

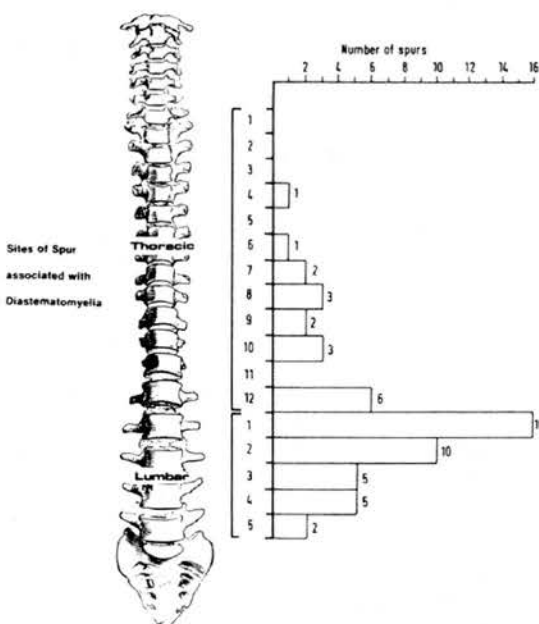


FIG. 2

Sites of spurs associated with diastematomyelia.

In addition to the spur, there also were bone abnormalities affecting the posterior elements of the spine, mainly spina bifida occulta. It occurred in thirty-eight of the forty-one patients with a diastematomyelia, and the laminar defect affected from two to ten adjacent vertebrae. The interpedicular distance in the affected vertebrae was also widened. The spur was situated near the center of the spina bifida in twenty seven patients and toward the upper end in eleven. The pedicles were also slightly thinner at the site of the diastematomyelia in twenty-four patients and the intervertebral discs were narrowed at the site of the spur in forty.

Nine patients had an intraspinal anomaly other than diastematomyelia, but only one of these anomalies could be identified on plain radiographs. This patient (Case 6) had a large teratoma that lay in the midline and extended from

the ninth thoracic to the fourth lumbar vertebra (Figs. 4 A, 4-B, and 4-C). In three other patients (Cases 5, 8, and 9) the intraspinal anomaly could not be specifically recognized on the plain radiographs but was indicated by a spina bifida occulta and widening of the spinal canal in the same region.

Myelographic Findings

The diastematomyelia always was clearly visible as a midline filling defect on the anteroposterior myelographic radiographs. The cleft or split in the dura extended over one to five vertebral levels (mean, three levels). The spur usually lay in the middle of the split in the dura, and in only three patients did it lie at the caudal end.

In five patients the myelogram also showed the presence of other intraspinal anomalies that appeared as filling defects, but their radiographic appearance usually was not specific for the anomaly. One defect was a neurenteric cyst (Case 1, Figs. 5-B and 5-C), another was a unilateral absence of nerve roots (Case 7), and three were intraspinal fibrous adhesions that distorted the column of contrast medium (Cases 4, 8, and 9).

In two patients (Cases 2 and 3) cysts could not be distinguished from the diastematomyelia with which they were closely associated.

Results of Surgical Treatment of the Intraspinal Anomalies

Thirty-four patients had an intraspinal spur or a cyst, or both, excised, and three had lysis of fibrous bands or adhesions. These operations were performed for a number of reasons. First, they were done to prevent the continuation of a progressive neural abnormality. Nine patients who were first seen before the age of five years had a progressive neural abnormality. Five patients (Cases 10 through 14) had a diastematomyelia, and a spur was excised at a mean age of 5.6 years (range, four to seven years). A sixth patient (Case 3) had a bone spur and a dermoid cyst removed from the same level at the age of four years and nine months. Prior to the operation three of these six patients had been incontinent of urine, but afterward they regained control of the bladder. The other preoperative neural deficits remained unchanged in all six patients, and they were followed for a mean of 6.1 years (range, four years and two months to eleven years and eight months) before spine fusion. The other three patients with a progressive neural deficit preoperatively had other types of intraspinal anomalies including one complex teratoma (Figs. 4-B and 4-C). This patient (Case 6) had a partial sensory recovery in one foot after operation and was followed to skeletal maturity without further neural deterioration. One patient (Case 9) who had fibrous adhesions released at the cauda equina had no change in neural status. A patient (Case 5) with a neurenteric cyst and lipofibromas had improvement of urinary incontinence, but during the nine years of follow-up before spine fusion there was no further change in the neural status.

These procedures were also done to prevent possible neural abnormalities in patients who were neurally normal.

Seven patients who were neurally normal had prophylactic surgery for an intraspinal anomaly before the age of five years (range, three months to four years and nine months). A diastematomyelic spur was removed from all seven patients and one (Case 1) also had a neurenteric cyst removed from a higher level (Figs. 5-B and 5-C). Following the operations there were no neural complications. The patients were followed for a mean of 7.8 years (range, three years to fourteen years and six months). None of these patients have as yet required a spine fusion.

Some procedures were performed to prevent possible neural deterioration in patients with a neural deficit who did not yet require correction and fusion. Seven patients had a neural deficit when they were first seen, and the intraspinal anomaly was treated surgically at a mean age of six years (range, one month to fourteen years). A diastematomyelic spur was removed from all seven patients, including one (Case 2) with an epidermoid cyst at the same level. After this operation there was no neural deterioration. Four pa-

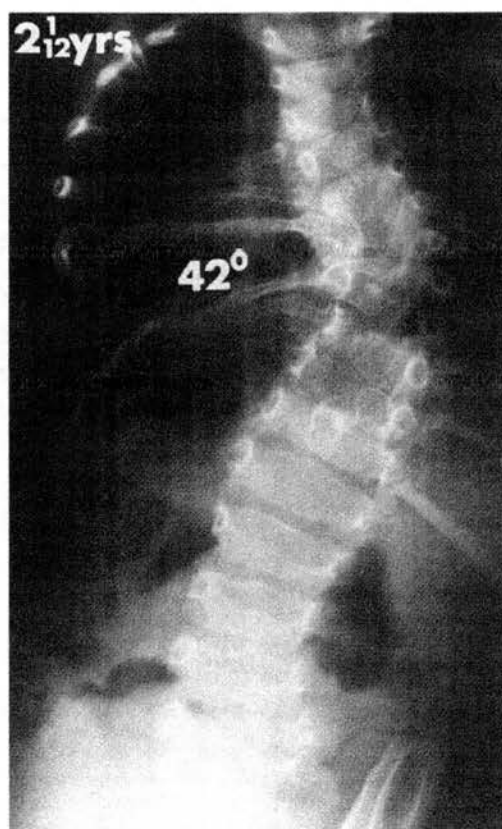


FIG. 3-A

Figs. 3-A, 3-B, and 3-C: Case 10.

Fig. 3-A: At the age of two years and one month, this girl had a 42-degree right thoracic scoliosis due to a unilateral unsegmented bar with contralateral hemivertebrae at the same level. The radiograph shows a midline bone spur arising from the twelfth thoracic vertebra, with a spina bifida occulta and widening of the spinal canal in this region.

tients were followed for a mean of 7.5 years before spine fusion (range, three years to fourteen years and five months). The other four patients did not have a spine fusion and were followed for a mean of 7.6 years (range, four to thirteen years). Two had reached skeletal maturity.

We also wanted to prevent neural complications during curve correction and spine fusion. In twelve patients the intraspinal anomaly was treated surgically prior to correction and spine fusion. This neurosurgical procedure was never performed at the same time as the spine fusion and usually preceded it by at least one month. Of the six patients who had normal neurological findings, five had a diastematomyelic spur removed and one (Case 4) had lysis of fibrous adhesions. These operations were performed between the ages of ten and fourteen years (mean age, 11.8 years), and there were no neural complications. The other six patients with abnormal neurological findings all had a diastematomyelic spur removed, and one (Case 8) also had lysis of fibrous adhesions. The neural deficit remained unchanged after these operations.

Correction of the Scoliosis and Spine Fusion

In thirty-two patients the scoliosis became sufficiently severe to require surgical treatment.

In fifteen patients a preoperative regimen of traction and a Risser plaster jacket was used. A few days later a

posterior spine fusion was performed with the patient in the jacket. In seven patients the intraspinal anomaly had been treated surgically as a separate procedure prior to this operation. In the other eight patients the anomalies had not been treated surgically. At the time of the posterior fusion the mean age of all fifteen patients was 10.7 years (range, nine years to thirteen years and eleven months). Before correction the curve measured a mean of 66 degrees (range, 33 to 102 degrees) and after surgery the curve had been corrected by a mean of 14 degrees (range, zero to 39 degrees). There was no change in the neural status of any of the patients: eleven had a neural deficit and four were normal. All had a solid fusion. They were followed for a mean of 5.7 years (range, two years and eleven months to ten years), during which time the neural status remained unchanged. Thirteen patients had reached skeletal maturity at the last follow-up.

In twelve patients the scoliosis was treated by Harrington instrumentation and posterior spine fusion (Fig. 3-C). The intraspinal anomaly had already been treated surgically as a separate procedure in all of these patients. At the time of posterior fusion the mean age was 11.8 years (range, nine years and two months to fifteen years and eight months). The curves before spine fusion measured a mean of 69 degrees (range, 35 to 99 degrees), and the Harrington instrumentation resulted in a mean correction of 21 degrees

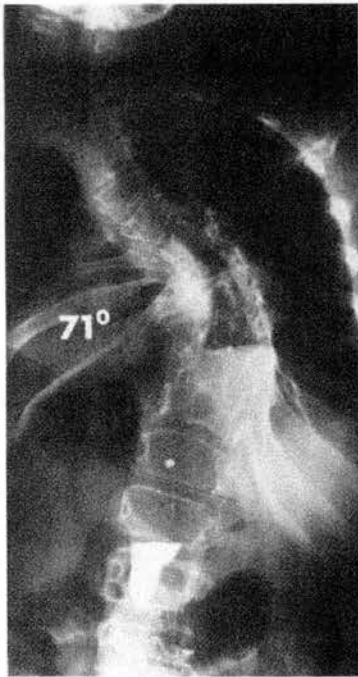


FIG. 3-B

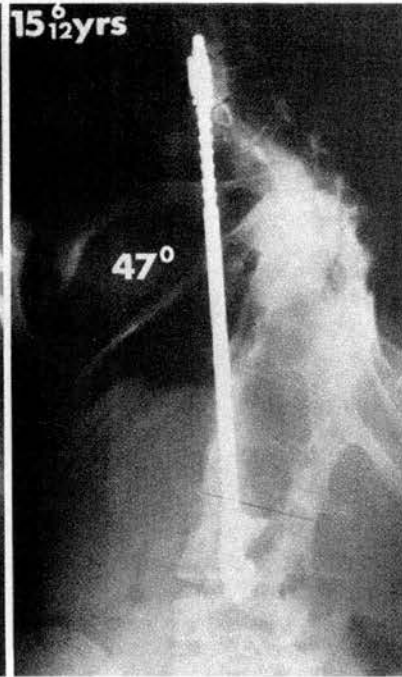


FIG. 3-C

Fig. 3-B: By the age of ten years the scoliosis had deteriorated to 71 degrees and required a spine fusion.

Fig. 3-C: The scoliosis was corrected to 45 degrees with Harrington instrumentation and there were no neural complications. At the age of fifteen years and six months the patient was skeletally mature and the spine was solidly fused, with minimum loss of correction. The neural deficit remained unchanged.

(range, 5 to 39 degrees). The intraoperative wake-up test was used in the majority of these patients. After surgery there was no change in the neural status of any of the patients: eight had a neural deficit and four were normal. All had a solid fusion. They were followed for a mean of 3.4 years (range, six months to seven years and five months), during which time the neural status remained unchanged. Three patients had reached skeletal maturity at the last follow-up.

Three patients had a very severe thoracolumbar curve due to a unilateral unsegmented bar with or without contralateral hemivertebrae. In these patients, after surgical treatment of the intraspinal anomaly we did an anterior vertebral-body resection or osteotomy as a second procedure, followed two to three weeks later by a posterior wedge resection and fusion with Harrington instrumentation. At the time of these operations the patients were in the sixth, thirteenth, and twenty-first years of life. Before correction the curves measured an average of 117 degrees (range, 80 to 148 degrees) and after the spine fusion they had been corrected by a mean of 49 degrees (range, 44 to 58 degrees). In one patient (Case 4) who previously had been neurally normal a partial paralysis of one lower limb developed following anterior resection of the bodies of the eleventh and twelfth thoracic vertebrae, which were unilaterally synostosed. No intraoperative correction of the scoliosis had been attempted. All of the muscles in one lower limb were Grade I in power or less and there was a very mild generalized diminution in sensation. Bladder control remained normal. This complication was thought to be due to an impairment of the blood supply to one-half of the bifid spinal cord at the level of the vertebral resection, which was also the site of a fibrous spur. There was no further neural deterioration following the second-stage correction with Harrington instrumentation, which was carried out three weeks later. Sensation rapidly returned to normal, and after one year of follow-up the motor power in the limb had recovered to at least Grade IV in all muscle groups, but there was generalized spasticity. There was no neural change after the operation in the other two patients, who were followed for more than three years after the fusion. One patient had a mild and the other (Case 9) had a severe neural abnormality affecting one lower limb.

In two patients the scoliosis was corrected by means of halo traction followed by posterior spine fusion and later by application of a Risser-Cotrel plaster jacket. The intraspinal bone spur had been excised from both patients before correction was attempted. At the time of surgery the patients were twelve and thirteen years old. Before correction the curves measured 25 and 75 degrees and after surgery they had been corrected by 5 and 23 degrees, respectively. After surgery the neural status of these patients remained unchanged. One had a mild and the other had a moderately severe unilateral neural abnormality affecting the lower limb. They were followed for more than three years without neurological change. Both had reached skeletal maturity.

Only four patients had no surgical treatment for either

the intraspinal anomaly or the scoliosis. Three of these patients had a diastematomyelia and one (Case 7) had a unilateral absence of nerve roots. They were followed for a mean of 6.7 years (range, two years and six months to fifteen years), without change in their neural status: three had a neural deficit and one was normal. When last seen, three had reached skeletal maturity and all four had a curve of less than 20 degrees.

Neural Abnormalities without Myelographic Evidence of an Intraspinal Anomaly

A unilateral neural deficit in the lower limb occurred in twelve of the 251 patients, but their myelograms showed no evidence of an intraspinal anomaly. In four of these twelve patients, a spina bifida occulta involved two or three adjacent vertebrae in the lumbar region, while in eight no radiographic abnormalities other than the congenital scoliosis were seen. All twelve patients had a very similar presentation and their clinical findings also were similar to those of the forty-six patients with an intraspinal anomaly. There were eleven girls and one boy.

Eight patients were seen because of a scoliosis, which was due to a unilateral unsegmented bar in six patients, to a block vertebra in one patient, and to complex anomalies in one patient. The site of the scoliosis was thoracolumbar

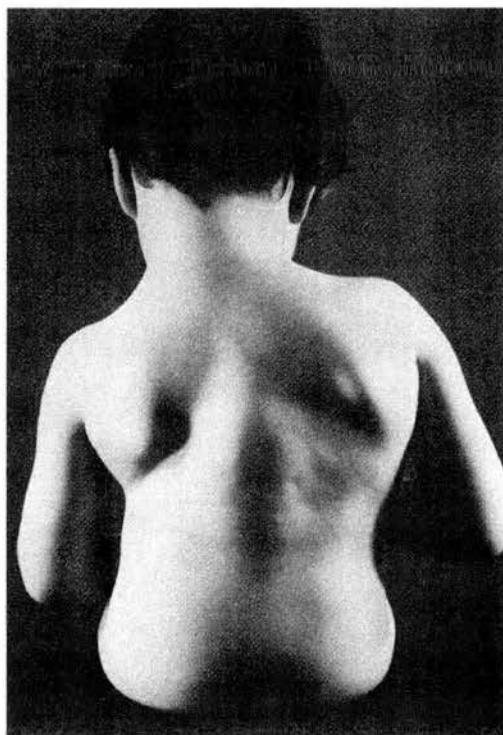


FIG. 4-A

Figs. 4-A, 4-B, and 4-C: Case 6.

Fig. 4-A: Clinical picture at the age of five years and ten months, showing a large lipoma overlying the spine.

in six patients, lower thoracic in one patient, and upper thoracic in one patient. When first seen the patients ranged in age from one year and six months to sixteen years old. A hairy patch overlying the spine was present in three patients. The ankle and knee reflexes were unilaterally absent or diminished in all of the patients and the Babinski sign was positive in two patients. Five patients had a sensory disturbance affecting part of the foot. There were no bladder

hemivertebra in one patient, to a wedge vertebra in one, to a lower thoracic unilateral unsegmented bar in one, and to multiple complex anomalies throughout the spine in one. A pigmented nevus overlying the spine occurred in one patient and a dermal sinus, in another. The ankle reflex was absent or diminished in all four patients and the knee reflex, in three patients. The Babinski sign was positive in two patients. All four patients had a sensory disturbance affecting



FIG. 4-B



FIG. 4-C

Fig. 4-B: The radiograph shows a right thoracic scoliosis due to complex anomalies. There is also a large fusiform enlargement of the spinal canal in the thoracolumbar region, with mottling of the posterior aspects of the vertebral bodies.

Fig. 4-C: The myelogram shows a large, smooth intraspinal lesion extending into the midline from the ninth thoracic to the fourth lumbar vertebra. At surgical exploration, this anomaly was found to be a complex teratoma.

disturbances. Varying degrees of muscle weakness distal to the knee resulted in pes cavus in five patients and clawing of the toes in the remaining three patients.

The other four patients were seen because of a foot deformity with a unilateral neural deficit and muscle imbalance. When first seen, these four patients ranged in age from eight months to ten years. At the time of diagnosis scoliosis was present in all of them; it was due to a lumbar

part of the foot. There were no bladder disturbances. Muscle weakness distal to the knee resulted in pes cavus, claw toes, and an inverted heel in all four patients.

The twelve patients in this group were followed for a mean of 7.6 years (range, two to fifteen years) after diagnosis, and eight had reached skeletal maturity when they were last seen. Six patients had correction of the scoliosis and spine fusion, without complications, between the ages

of eight and thirteen years. During the period of follow-up there was no change in the neural status of any of the patients.

Discussion

This study has shown that congenital intraspinal anomalies found in association with congenital scoliosis are much more common than previously reported. An intraspinal anomaly was diagnosed in forty-six of 251 patients with congenital scoliosis, an incidence of 18.3 per cent. I also found that an additional twelve (4.8 per cent) of the 251 patients with congenital scoliosis had a unilateral neural deficit in the lower limbs; these were similar to those found in the patients with intraspinal anomalies, but the patients had no myelographic evidence of an anomaly.

A diastematomyelia was the commonest anomaly and occurred in forty-one (16 per cent) of the 251 patients. In addition to the diastematomyelia, four of these patients had other types of intraspinal anomalies, while five patients (2 per cent) did not have a diastematomyelia but did have other types of intraspinal anomalies. These included neurenteric cysts, dermoid cysts, epidermoid cysts, teratomas, lipomas, absence of nerve roots, fibrous bands, and a tight filum terminale. Myelograms were not made for all of the patients with congenital scoliosis. The prevalence of occult intraspinal anomalies could therefore be even higher than the 18 per cent that was recorded. In the only reported series that was comparable to this one, Winter et al.¹¹ found a diastematomyelia in only 4.9 per cent of 392 patients with congenital scoliosis, but other types of intraspinal anomalies were not studied and myelography was not performed on all patients. Gillespie et al. reported on thirty-one patients who had an intraspinal anomaly associated with congenital scoliosis; seventeen of them had a diastematomyelia and fifteen had developmental abnormalities that were similar to those found in the present series. These authors did not, however, report the over-all prevalence of these anomalies in their patients with congenital scoliosis. It therefore may be concluded that the incidence of intraspinal anomalies in children with congenital scoliosis is probably as high as 20 per cent or more.

Keim and Greene and Hood et al., in studies of patients with diastematomyelia, found that 60 to 70 per cent had congenital scoliosis. A scoliosis was not present in four of the forty-one patients with diastematomyelia in the present series at the time when the intraspinal anomaly was first diagnosed, but it had developed in all patients by the age of eight years. The intraspinal anomalies were associated with all sites and types of congenital scoliosis. The most commonly associated osseous anomaly in the spine causing the congenital scoliosis was a unilateral unsegmented bar with contralateral hemivertebrae occurring in the lower thoracic or thoracolumbar region (Table II). Twenty-one patients had this pattern of scoliosis and eleven of them (52 per cent) had an intraspinal anomaly.

Of the 251 patients with congenital scoliosis, girls were more commonly affected than boys (ratio, 2.5:1), but of the

forty-five patients who also had an intraspinal anomaly, girls were affected even more often (ratio, nearly 7:1).

Clinically the intraspinal anomalies presented in several ways. Fifteen (33 per cent) of the forty-six patients, all of whom were less than five years old, were seen not because of their scoliosis but because of either a cutaneous abnormality on the back or a neural deficit in the lower limbs or the bladder. The other thirty-one patients were seen because of the scoliosis. An intraspinal anomaly was first suspected in nineteen patients because of abnormal neurological signs. Twelve patients had no such signs, and the intraspinal anomaly was suspected in seven of them because of the abnormality on the plain radiographs of the spine. In the remaining five patients, the intraspinal anomaly was detected only when routine myelography was performed prior to surgery for the scoliosis.

Retrospectively a bone spur could be identified on the early radiographs of thirty-three of the forty-one patients with diastematomyelia. However, the spur became increasingly more difficult to recognize as the scoliosis worsened and the spine became more distorted. When there was a fibrocartilaginous spur (present in four patients) it could only be demonstrated myelographically. In twenty-one patients

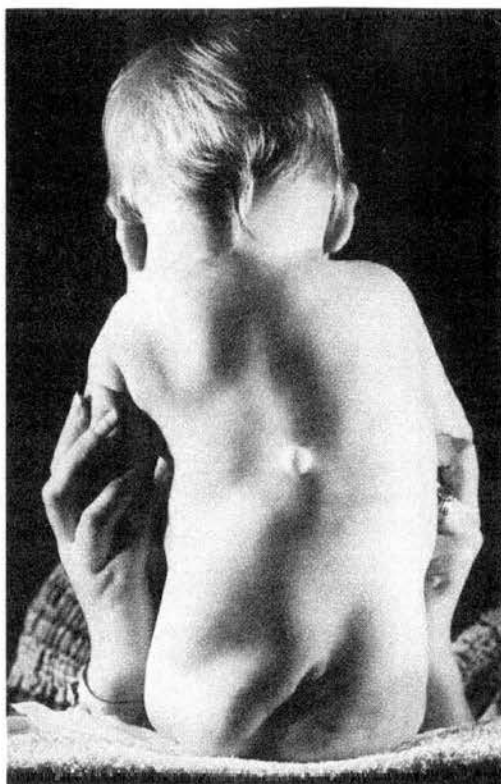


FIG. 5-A

Figs. 5-A, 5-B, and 5-C: Case 1.

Fig. 5-A: This eight-month-old girl had a small subcutaneous lipoma overlying the spine.

(50 per cent) the osseous or fibrocartilaginous spur was at the same level as the abnormalities in the vertebral bodies that produced the scoliosis, and that level most frequently was between the twelfth thoracic and fourth lumbar vertebrae (Fig. 2). The commonest site was at the first lumbar vertebra (sixteen patients, or 39 per cent). A spina bifida occulta with widening of the interpedicular distance and narrowing of the disc spaces was nearly always present at the site of the diastematomyelia. The combination of these anomalies in a patient with congenital scoliosis should, therefore, make one very suspicious of an intraspinal anomaly which may not be apparent on plain radiographs of the spine. Of the other types of intraspinal anomalies, only a teratoma (seen in one patient) could be detected on the plain radiograph of the spine (Fig. 4-B).

Occult intraspinal anomalies were also frequently associated with an abnormality of the skin or subcutaneous tissues overlying the spine. A hairy patch, an area of telangiectasis, or a subcutaneous lipoma was present in 74 per

cent of the patients with intraspinal anomalies of all types. In all but four of these patients the superficial abnormality overlay the spine in the same region as the intraspinal anomaly. These stigmata did not, however, always indicate a structural intraspinal anomaly, because similar abnormalities occurred in five patients who, although having a neural deficit, did not have myelographic evidence of an anomaly.

James and Lassman have described at length the clinical syndromes in which the lower limbs of children show abnormalities associated with occult congenital intraspinal anomalies of all types. The common findings are a generalized mild underdevelopment and shortening of one lower limb and foot associated with neural abnormalities and a paralytic foot deformity which is often progressive. Bowel and bladder function may be disturbed and occasionally both lower limbs are affected. These findings, associated with a neural abnormality, occurred to some degree in thirty (65 per cent) of the patients with an intraspinal anomaly in this series, and eight additional patients (17 per cent) with an



FIG. 5-B



FIG. 5-C

Fig. 5-B: The radiograph shows a right thoracolumbar scoliosis due to a unilateral unsegmented bar with a contralateral hemivertebra at the same level. A midline bone spur is shown arising from the first and second lumbar vertebrae, with a spina bifida occulta and widening of the spinal canal in the same region.

Fig. 5-C: The myelogram shows that in addition to the diastematomyelia in the lumbar region there is a smooth, round intraspinal anomaly in the upper thoracic region. At surgical exploration the upper thoracic anomaly was found to be a neurenteric cyst.

intraspinal anomaly had mild underdevelopment and shortening of one lower limb and foot but were neurally normal. The remaining eight patients (17 per cent) had no abnormalities in the lower limbs despite the presence of an intraspinal anomaly. Although the neural abnormalities were not noted to progress after the age of six years, the foot deformity often did progress due to continued growth and abnormal use of the limb in the presence of muscle imbalance. This progressive deformity of the foot can be mistakenly attributed to further neural deterioration.

It has been suggested that the neural abnormalities that occur in association with a diastematomyelia and other congenital intraspinal anomalies are due to traction on the spinal cord. In early embryonic life, the spinal cord occupies the entire length of the vertebral canal, but due to differing rates of growth and differentiation of the neural tube compared with the osseous elements there is an apparent cranial migration of the conus medullaris until it reaches its final level at the upper border of the second lumbar vertebra. During this process, any tethering of the cord by an intraspinal anomaly could result in a neural deficit. Although this hypothesis is attractive, it does not fit well with the facts. Barson has shown that the ascent of the spinal cord is most rapid during the first half of intra-uterine life, and that the final level is reached nine weeks after birth. Neural abnormalities due to simple axial tethering of the cord ought to be present in most children at birth and little, if any, deterioration would ordinarily be expected thereafter. Secondly, one would expect that axial traction on the cord would produce bilateral abnormalities in the lower limbs, whereas in the majority of patients only one lower limb was affected. Thirdly, if the spur itself were to significantly tether the cord, one would expect it to lie at the most caudal end of the split in the dura, whereas in this series the myelograms showed the spur to lie in that position in only three patients.

Guthkelch has suggested an alternative and more acceptable theory, that the unilateral syndrome affecting the lower limbs of these children may be due to a unilateral developmental anomaly of the cord on one side of the diastematomyelia. On the other side, the cord may be normally developed and hence the corresponding limb would be normal. The resulting neural abnormality is present at birth but is not usually recognized until it is very severe. It is only when the child begins to walk and becomes more mobile that the neural deficit becomes apparent, especially if a paralytic deformity of the foot develops. Some evidence to support this theory was found in the twelve patients in this series who, although they had normal myelograms, had unilateral neural abnormalities similar to those found in the patients with a proved structural intraspinal anomaly. It is possible that these patients with normal myelograms had a unilateral anomaly of the cord, although there was no structural bifurcation. Shaw suggested "myelodysplastic syndrome" as a suitable description of the characteristic abnormalities in the lower limbs of these children.

Neural deterioration occurred in five of the children

with a diastematomyelia who were seen before the age of six years. There was no neural deterioration in any of the other children with a diastematomyelia who were seen after the age of six years. The neural abnormality in the patients who had normal myelograms did not change, as one would expect with a unilateral anomaly of the cord. In this series, it appears that if neural worsening had not occurred before the age of six years it was unlikely to occur thereafter. Excision of the spur improved bladder control in the three patients who were incontinent but did not relieve the other neural abnormalities, although there was no further worsening of a neural deficit in any of the patients. Neither the severity of the scoliosis nor its rate of deterioration had any association with the progression of a neural abnormality.

It is widely accepted that there are two good indications for surgical treatment of a structural intraspinal anomaly. One is a progressive neural abnormality and the other is prophylaxis before attempting to correct an associated scoliosis. In my opinion, once diagnosed, an intraspinal anomaly should be treated as a prophylactic measure in all children who are less than six years old who have congenital scoliosis, regardless of their neural status. The intraspinal anomaly should also be treated immediately in patients who are more than six years old if the congenital scoliosis is thought to be likely to require correction and fusion at a later stage. Routine myelography should be performed on all patients with congenital scoliosis who have clinical or radiographic evidence that is indicative of an anomaly, and on patients who are about to undergo corrective spine surgery. A plain radiograph, even if it shows an osseous intraspinal spur, is not sufficient, because other types of intraspinal anomalies may also be present and can only be identified by myelography or computerized tomography. Excision of a spur or cyst, or lysis of adhesions, rarely improved a neural deficit in my series, but it probably will prevent deterioration and decrease the risk of traction on the cord if the scoliosis is to be corrected and fused. None of the patients whose intraspinal anomaly was treated surgically had neural complications, despite the use of Harrington instrumentation to correct the scoliosis (Figs. 3-C and 3-D), and neither did the five patients with non-progressive neural abnormalities or the three with no neural deficit who did not have a diastematomyelic spur removed prior to surgical correction of the scoliosis and fusion of the spine in a Risser localizer jacket. The fact that this procedure was carried out without neural complications may have been due to the relatively small degree of correction of the scoliosis that was obtained. I strongly recommend, however, that all intraspinal anomalies be treated surgically before any more effective method of correction of scoliosis, such as Harrington instrumentation, is applied, particularly if the patient is to be anesthetized during the correction. The intraoperative wake-up test should also be used during the correction to detect any possible neural deterioration.

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HEMIVERTEBRA AS A CAUSE OF SCOLIOSIS

A STUDY OF 104 PATIENTS

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From the Edinburgh Scoliosis Unit

We studied 104 patients with a total of 154 hemivertebrae which had produced scoliotic curves. Of the hemivertebrae 65% were of a fully segmented (non-incarcerated) type, 22% were semi-segmented and 12% were incarcerated.

We found that the degree of scoliosis produced depended on four factors: first, the type of the hemivertebra; secondly, its site; thirdly, the number of hemivertebrae and their relationship to each other; and finally, the age of the patient. Semi-segmented and incarcerated hemivertebrae usually do not require treatment. Fully segmented non-incarcerated hemivertebrae may require prophylactic treatment to prevent significant deformity.

A hemivertebra due to complete unilateral failure of formation may result in scoliosis, but its severity varies greatly and there is controversy regarding treatment. Opinions range from total excision to total neglect of the hemivertebra (Moe et al. 1978). To rationalise management a better understanding of the natural history of the different types and sites of hemivertebrae is needed.

This paper does not aim to recommend any specific treatment but to provide an indication of the need for and the timing of prophylactic treatment before there is significant deformity, thereby helping to avoid unnecessary treatment.

MATERIAL AND METHODS

All patients with radiographic evidence of hemivertebrae seen at the Edinburgh Scoliosis Clinic between 1958 and 1984 were reviewed and those with one or more scoliotic curves, each directly attributable to one or more hemivertebrae, were studied. Patients with hemivertebrae associated with a unilateral unsegmented bar (Nasca, Stelling and Steel 1975) were excluded as their prognosis relates more to the bar. Patients whose hemivertebra caused a kyphosis or a kyphoscoliosis in which kyphosis was the predominant deformity were also excluded because their natural history is very different from those

in which there is scoliosis alone. This left 104 patients with hemivertebrae producing a true congenital scoliosis. There were 66 females and 38 males those ages at diagnosis ranged from birth to 20 years. When first seen, four patients were skeletally mature; the remaining 100 were followed without treatment for a mean of 5 years (range 6 months to 15 years 2 months). At their last clinic visit, 73 patients had had no treatment and 33 of them had reached skeletal maturity. The remaining 31 patients had been followed without treatment for a mean of 3 years 8 months before eventually being treated either in a brace or by spinal fusion.

The types and sites of the hemivertebrae were diagnosed by reviewing the anteroposterior radiographs. The scoliotic curves were measured using the Cobb method, on radiographs taken with the patient standing, taking care that measurements were made from the same spinal levels on all the serial radiographs of each patient. Skeletal maturity was diagnosed when there was complete ossification and fusion of the iliac apophyses. Pelvic obliquity, decompensation or listing of the trunk, elevation of the shoulder and tilting of the head were assessed on the basis of the spinal radiographs and clinical photographs made with the patient standing.

Sites and types of hemivertebra

The 104 patients had a total of 154 hemivertebrae which were divided into three groups based on the pathological anatomy of the hemivertebra: fully segmented (non-incarcerated), semi-segmented and incarcerated. For each patient, the vertebrae in the thoracic and lumbar regions were numbered on the radiograph in a cranial to caudal direction, counting each vertebra and each hemivertebra as one unit. A thoracic vertebra or hemivertebra was defined by the attachment of a rib or ribs. Using this

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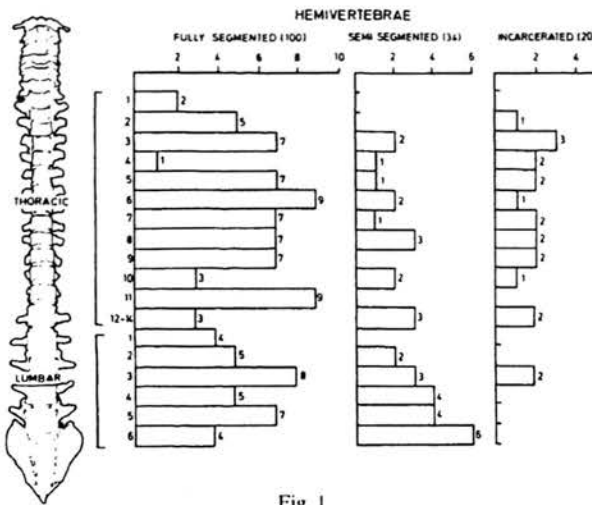


Fig. 1

The sites and types of 154 hemivertebra in 104 patients.

method 34 patients had 13 thoracic vertebrae and six patients had 14, while 34 patients had six lumbar vertebrae. The sites of the various types of hemivertebrae are shown in Figure 1. In all, 70 patients (67%) had a single hemivertebra, 28 (27%) had two hemivertebrae and six patients (6%) had more than two hemivertebrae. Multiple hemivertebrae were never adjacent, always being separated by at least one relatively normal vertebra.

Fully segmented (non-incarcerated) hemivertebrae. Of the 154 hemivertebrae, 100 (65%) were fully segmented and non-incarcerated, and were fairly evenly distributed throughout the spine (Fig. 1) with equal numbers on each side.

Fully segmented hemivertebrae were usually triangular in shape with the disc spaces above and below appearing to be relatively normal. The hemivertebra was wedged between two relatively normal vertebrae which

often became slightly wedge-shaped during growth but the disc spaces were preserved (Fig. 2). The lateral margin of the hemivertebra was equal or nearly equal in height to the adjacent vertebrae and 23 of these 100 hemivertebrae extended across the midline. All had a single pedicle and in the thoracic region they were associated with a rib; this could result in an extra rib on one side of the spine. A single unilateral hemivertebra always lay at the apex of a definite scoliosis and a characteristic of this type was that, as the scoliosis deteriorated, the body of the hemivertebra tended to protrude slightly from the lateral margin of the spine (Fig. 2).

Semi-segmented (non-incarcerated) hemivertebrae. Thirty-four (22%) of the hemivertebrae were semi-segmented. They were commonest in the lumbar region (Fig. 1) and tended to be single.

Semi-segmented hemivertebrae were similar in shape to the fully segmented type but the body of the hemivertebra was synostosed with one of its neighbouring vertebra, with no intervening disc space (see Fig. 17). The other disc space either above or below the hemivertebra was relatively normal. Of these 34 hemivertebrae, 19 were synostosed with the vertebra above and 16 with the vertebra below. Three cases of hemivertebra, diagnosed in the first year, initially appeared to be fully segmented but during later ossification of the spine it became apparent that they were semi-segmented.

Incarcerated hemivertebrae. The 20 incarcerated hemivertebrae (13%) formed the least common type. They were most frequent in the thoracic region (Fig. 1) and tended to be single.

An incarcerated hemivertebra was usually more ovoid in shape and smaller than a fully segmented one. It was tucked into the spine and set in a niche scalloped out of the adjacent vertebrae (see Fig. 18). The vertebrae above and below were shaped in such a way that they tended to compensate for the hemivertebra and as a

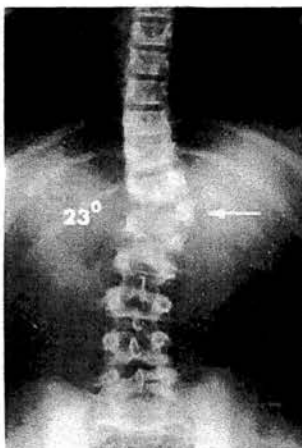


Fig. 2

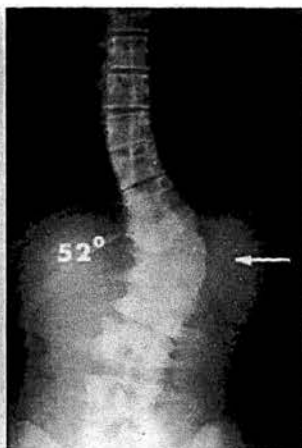


Fig. 3



Fig. 4

Case 1. Figure 2 A boy aged 11 years with a 23° right thoracolumbar scoliosis due to a fully segmented (non-incarcerated) hemivertebra at L1. Figure 3 - No treatment was given and by the age of 20 years the patient was skeletally mature and the curve had deteriorated to 52°. Figure 4 - Despite the severity of the curve the cosmetic deformity is relatively mild.

result the general alignment of the spine remained straight with minimal scoliosis. The disc spaces above and below the incarcerated hemivertebra were often narrow and sometimes poorly formed. The lateral margin of the hemivertebra was usually two-thirds or less than the height of the adjacent relatively normal vertebrae and the hemivertebra did not extend across the midline. All the incarcerated hemivertebrae had a single pedicle and in the thoracic region they were all associated with a rib.

Rib and chest anomalies. Congenital rib fusions were present in 10 of the 104 patients and were unilateral or bilateral, affecting two or three adjacent ribs at a site distant from the vertebral column. They were not thought to affect the development of the scoliosis; the site of the fusion was not specifically related to the site or side of the hemivertebra.

A chest wall defect was present in four patients and was due to the absence of several adjacent ribs, always on the convex side of the scoliosis and just below the level of the hemivertebra. Congenital elevation of the scapula (Sprengel's shoulder) was present in two patients who had hemivertebrae at T1 or T2. Unfortunately, in both patients, both abnormalities were on the same side and combined to increase the deformity produced by the elevated shoulder.

Other congenital anomalies. Occult intraspinal anomalies were diagnosed by myelogram in seven patients (7%); six had diastematomyelia and one a unilateral absence of lumbar nerve roots. Four of these patients had hemivertebrae at the lumbosacral junction.

Other congenital anomalies were also common: Klippel Feil syndrome and heart anomalies were each seen in six patients; thumb deformity and foot deformity each in five patients; rectal atresia, urinary anomalies, radial club hand, or congenital dislocation or subluxation of the hip were seen in four patients. Goldenhar syndrome was seen in three patients; facial asymmetry and tracheo-oesophageal fistula in two patients each; while cleft palate and reduction of the lower limb were each present in one patient of the series.

RESULTS

Of the 104 patients, 87 had a single congenital scoliosis; 70 were due to a single hemivertebra (67%) and 17 to two hemivertebrae on the same side (16%). Eleven patients had two congenital curves (11%) each of which was due to a single hemivertebra on opposite sides of the spine. Six patients had multiple congenital curves (6%) associated with three or more hemivertebrae which alternated on either side of the spine.

For the purpose of this study the scoliotic curves were grouped according to the type of hemivertebra and then subdivided by the level of the hemivertebra or, if there were two unilateral hemivertebrae, the apex of the curve. The rate of deterioration of the untreated scoliotic

Tables I, II, III and IV shows the mean yearly rate of deterioration in degrees of scoliotic curves in untreated patients due to different types of hemivertebra occurring in different regions of the spine, both before and after 10 years of age.

Table I. Scoliosis due to a single fully segmented (non-incarcerated) hemivertebra

Site of hemivertebra	Number of patients	Rate of deterioration (degrees per year)	
		Before 10 years	After 10 years
T1 T4	4	1	1.5
T5 L1	18	1.4	3
L2 L4	9	0.7	1.7
L5	9		

Table II. Scoliosis due to two unilateral fully segmented (non-incarcerated) hemivertebrae

Apex of scoliosis	Number of patients	Rate of deterioration (degrees per year)	
		Before 10 years	After 10 years
T1 T4	4	1.5	2.25
T5 L1	13	3	3.2

Table III. Scoliosis due to a single semi-segmented hemivertebra

Site of hemivertebra	Number of patients	Rate of deterioration (degrees per year)	
		Before 10 years	After 10 years
T1 T4	2	0	0
T5 L1	6	0	<1
L2 L4	6	0	1
L5	5		

Table IV. Scoliosis due to a single incarcerated hemivertebra

Site of hemivertebra	Number of patients	Rate of deterioration (degrees per year)	
		Before 10 years	After 10 years
T1 T4	3	0	0
T5 L1	7	0	<1
L2 L4	1	0	<1

curves was calculated for each type of hemivertebra and site of curve, both before the age of 10 years and also after 10 years, during the adolescent growth spurt (Tables I, II, III and IV).

Fully segmented (non-incarcerated) hemivertebrae

Single hemivertebra. Forty patients had a single vertebra of this type producing a single congenital scoliosis. These hemivertebrae were considered in four regions of the spine each of which gave different problems and prognoses. Follow-up without treatment was for a mean of 5 years 10 months (range 1 to 13 years).

T1 to T4. Four patients had hemivertebrae in this region; three at T3 and one at T2. Three of these patients were seen before the age of 10 years when the mean rate of deterioration of their curves was 1° per year (range 0.7 to 1.1°). At the age of 10 years their mean curvature was 32° (range 28° to 39°). Two of these patients and a third, diagnosed later, were followed after the age of 10 years; the rate of deterioration increased slightly to 1.5° per year (range 1° to 2°). Although these curves deteriorated relatively slowly and never became very large, they caused a cosmetic deformity by elevation of one shoulder and in one patient tilting of the head to the opposite side. To control deformity, two patients had spinal fusion at the ages of 11 and 15 years when their curves measured 32° and 35° respectively, while the third patient reached skeletal maturity untreated with a 48° curve.

T5 to L1. Of 18 patients with hemivertebrae in this region of the spine (Figs 2, 3 and 4), eight were seen before the age of 10 years and followed without treatment; their mean rate of deterioration was 1.4° per year (range 1° to 2.3°). Four of these patients were last reviewed at a mean age of 6 years 1 month (range 3 years 4 months to 7 years 2 months), when their mean curvature was 32° (range 23° to 37°). The other four patients were followed to 10 years when their mean curvature was 41° (range 31° to 47°).

After the age of 10 years, 12 patients were followed without treatment and the mean rate of deterioration of the curves increased to 3° per year (range 2.5° to 4.5°). Three patients eventually had spinal fusion at a mean age of 12 years 10 months (range 12 to 15 years) when their mean curvature was 51° (range 44° to 58°). Nine patients reached skeletal maturity without treatment when their mean curvature was 46° (range 35° to 58°).

These lower thoracic and thoracolumbar curves produced only mild to moderate cosmetic deformity because the vertebrae were not significantly rotated and there was only a small rib hump (Fig. 4). Only two of the 18 curves were decompensated, causing the patient to list slightly to one side. An additional problem associated with two of the curves due to hemivertebrae at T6 was the development of a long compensatory thoracolumbar curve which later became fixed. This *secondary structural curve*, containing no congenital anomaly, was much more rotated than the primary congenital curve; it produced a rib hump which was the main cosmetic deformity. Both of these curves were first diagnosed after the age of 10 years and followed to skeletal maturity without treatment, when the thoracic congenital curves measured 44° and 50° and the structural compensatory curves 38° and 52° respectively.

L2 to L4. Nine patients had a single fully segmented hemivertebra in this region (Figs 5 and 6). Eight of these were seen before the age of 10 years when the mean rate of deterioration of their curves was 0.7° per year (range 0° to 1.4°). One patient was last seen at two years of age with a 24° curve, while a second patient, last seen at 6 years 8 months, had a 35° curve. The other six patients

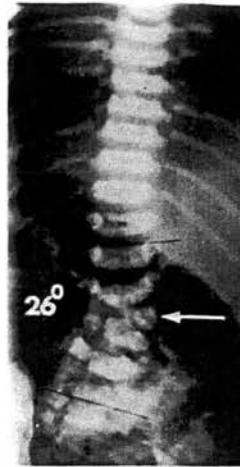


Fig. 5

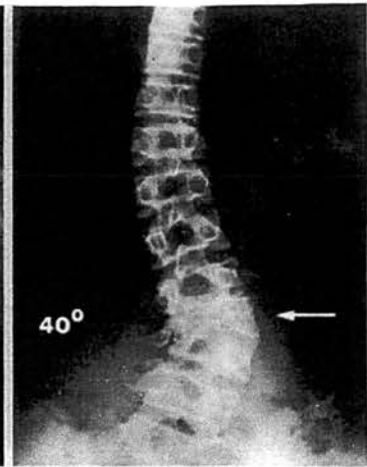


Fig. 6

Case 2. Figure 5 - A one-day-old girl with a 26° right lumbar scoliosis due to a fully segmented (non-incarcerated) hemivertebra at L4. Figure 6 - No treatment was given and by the age of 13 years the curve had deteriorated to 40°.

had been followed untreated to 10 years of age when their mean curvature was 33° (range 22° to 43°). Five of these patients and one other were followed untreated after the age of 10 years when the mean rate of deterioration of their curves increased to 1.7° per year (range 1° to 2.3°). Three patients were last seen between the ages of 11 and 14 years when their mean curvature was 42° (range 40° to 46°). The three patients who had been followed to skeletal maturity had a mean curvature of 45° (range 40° to 55°).

These lumbar curves produced only a relatively mild cosmetic deformity because they were not significantly rotated and there was no rib hump. None of the curves were decompensated and there was no listing of the trunk. The pelvis remained level in all but three patients who had mild leg-length discrepancy.

Lumbosacral. Nine patients with a single fully segmented hemivertebra at the lumbosacral junction showed a very short congenital scoliosis from L4 or L5 vertebra to the sacrum (Figs 7, 8 and 9). Three patients had six lumbar vertebrae and two had four lumbar vertebrae, including the hemivertebra.

The lumbosacral hemivertebra caused the lumbar spine to take off obliquely from the sacrum and, in order to compensate, all the patients developed a long secondary thoracolumbar or lumbar curve which was initially mobile but later became fixed. This *secondary structural curve*, which contained no congenital anomalies, extended upwards to T10, T11 or T12 and was always much larger and more rotated than the primary congenital curve. This secondary curve did not fully compensate for the congenital scoliosis and as a result the upper part of the body listed to the side opposite the hemivertebra in the six patients who had a level pelvis (Fig. 9). In three patients a leg-length discrepancy fortunately produced a compensatory pelvic obliquity, allowing the upper body to remain balanced.

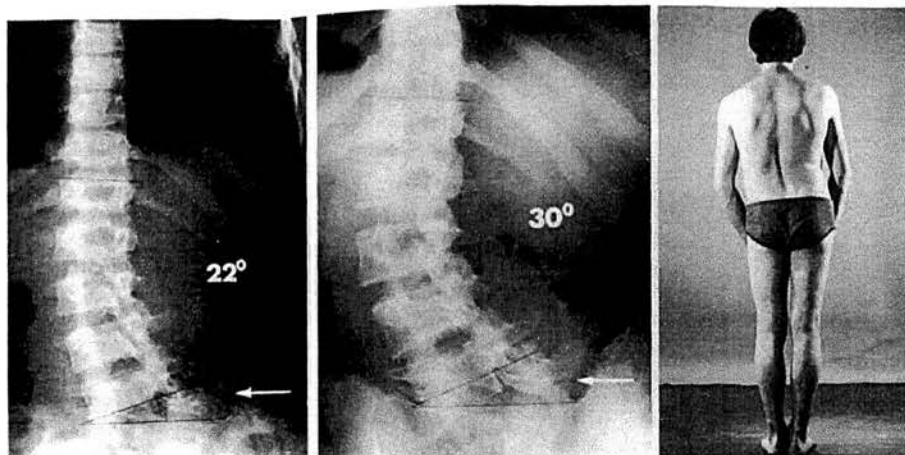


Fig. 7

Fig. 8

Fig. 9

Case 3. Figure 7 A boy aged 12 years 8 months with a single fully segmented (non-incarcerated) hemivertebra at the lumbosacral junction. This causes the lumbar spine to take off obliquely from the sacrum and a long secondary lumbar curve has developed which measures 22° and does not contain any congenital anomalies. Figure 8 No treatment was given and by the age of 17 years the secondary lumbar curve has deteriorated to 30°, and become fixed and rotated. The pelvis remains level. Figure 9 The main cosmetic deformity is due to the secondary lumbar curve and the tendency to list to the left.

Eight of the nine secondary structural curves behaved in a very similar manner. Only two of these patients were seen before the age of 10 years and neither of their curves exceeded 40°. After the age of 10, the eight secondary structural curves deteriorated at a mean rate of 3° per year (range 2° to 4°). Five patients reached skeletal maturity without treatment with a mean curvature of 44° (range 33° to 54°); all had a significant cosmetic deformity (Fig. 9). Two patients had spinal fusion at the age of 13 years 6 months when they both had secondary structural curves measuring 50°. The eighth patient was last seen untreated at 13 years when she had a 40° secondary curve. The ninth patient had a much more malignant deformity; she was first seen at the age of 3 years 5 months with a 42° secondary curve which deteriorated at 3° per year up to the age of 10 years and at 7° per year thereafter. This curve reached 100° at the age of 13 years 5 months resulting in a very severe deformity.

Double unilateral hemivertebrae. Seventeen patients had two fully segmented hemivertebrae on the same side of the spine. The two hemivertebrae were separated by from 1 to 4 relatively normal vertebrae (mean 2). The patients were followed without treatment for a mean of 3 years 3 months (range 6 months to 12 years).

T1 to T4. Four patients with two unilateral hemivertebrae each had single curves whose apex lay between T1 and T4 inclusive (Figs 10 and 11). Three of these patients were seen before the age of 10 when the mean rate of deterioration was 1.5° per year (range 1° to 2°). By the age of 7 years 8 months, one patient had developed a 43° curve which caused a significant cosmetic deformity, by elevating the shoulder and tilting the head (Fig. 11), and needed treatment. Two patients reached 10 years without treatment when they had curves of 47° and 40°; these also caused significant deformity from an elevated shoulder.

After the age of 10 years, the rate of deterioration of these curves increased to 2° and 2.5° per year; both were treated by spinal fusion at the ages of 12 and 13 years for curves measuring 51° and 48° respectively.

T5 to T12. Thirteen patients had two unilateral hemivertebrae with a single curve with apex from T5 to T12 (Fig. 12). Before the age of 10 years, 10 of these patients showed a mean rate of deterioration of 3° per year (range 1.5° to 4°). Seven of these were eventually treated in a brace at a mean age of 3 years 5 months (range 2 years 2 months to 4 years 10 months), when their mean curve was 53° (range 43° to 74°). The other three patients in this subgroup were last reviewed at a mean age of 9 years 6 months (range 8 to 10 years) when their mean curvature was 50° (range 43° to 55°). After the age of 10 years,

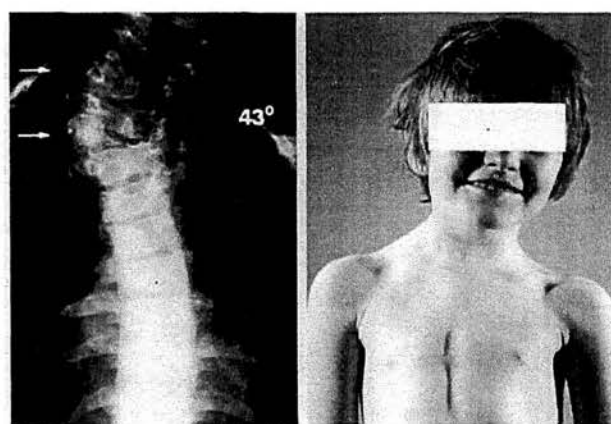


Fig. 10

Fig. 11

Case 4. Figure 10 - A girl aged 7 years 8 months with a 43° right upper thoracic scoliosis due to two unilateral fully segmented (non-incarcerated) hemivertebrae at T2 and T4. Figure 11 Her cosmetic deformity is due to elevation of the right shoulder and slight tilting of the head to the left.

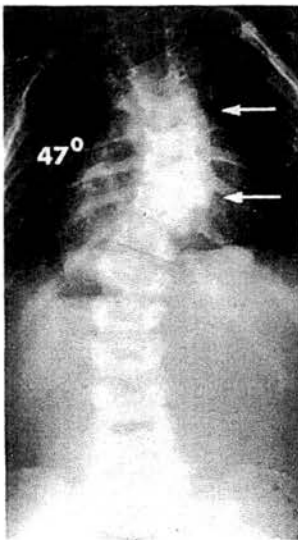


Fig. 12



Fig. 13

Cases 5 and 6. Figure 12 - A boy aged 4 years 10 months with a 47 right thoracic scoliosis due to two unilateral fully segmented (non-incarcerated) hemivertebrae at T6 and T9. Figure 13 - A skeletally mature girl aged 18 years with two opposing hemivertebrae in the same region at T4 and T6. These hemivertebrae produce curves which balance each other and cause minimal cosmetic deformity.

four patients followed without treatment showed an increased mean rate of deterioration of 3.2 per year (range 2.5 to 4). Three of these patients had a spinal fusion at a mean age of 14 years 7 months (range 12 years 3 months to 16 years 2 months), when their mean curve was 60 (range 57 to 62); the fourth was untreated at skeletal maturity when his curve was 72.

All these patients developed a significant cosmetic deformity before the age of five years, due to the severity of their curve and the presence of a rib hump. In addition, 6 of the 13 patients had decompensated curves and listed to one side. Two patients with curves whose apex was at T6 and T7 developed secondary structural curves in the thoracolumbar region. These two patients were untreated at age 16; their congenital curves measured 50 and 60 and their structural compensatory curves were 82 and 77 respectively.

Double opposing hemivertebrae. Eleven patients had two fully segmented hemivertebrae on opposite sides of the spine, resulting in double congenital scoliosis. These patients could be divided between three subgroups, depending on the sites and relationship of the hemivertebrae; each had a different prognosis.

1. Four patients had two opposing hemivertebrae occurring in the *same region* within one or two segments of each other, three in the thoracic spine and one in the lumbar region. These pairs of hemivertebrae caused small kinks in the spine which never became large and balanced each other to produce minimal cosmetic deformity at maturity (Fig. 13).

2. Two patients had opposing hemivertebrae that were more widely separated and in *different regions* of the spine. These hemivertebrae produced much bigger

curves, similar to those produced by single hemivertebrae in the same region. The resulting curves were unbalanced and caused listing of the upper trunk to one side. One patient had opposing lower thoracic and lumbar curves which measured 54 and 61 at the age of 10 years and required treatment because of decompensation of the trunk (Figs 14 and 15). The second patient had opposing lower thoracic and thoracolumbar curves which measured 48 and 49 respectively at the age of 12 years 6 months. This patient was balanced by slight shortening of one leg which compensated for the tendency to list.

3. Five patients had the combination of a *lumbosacral hemivertebra* with a *second hemivertebra on the opposite side in the lumbar region* (L1, L2 and three at L3). These patients were all unbalanced, listing to the side opposite the lumbosacral hemivertebra. Four of the patients were followed without treatment to skeletal maturity. Their lumbar curves measured 50, 54, 55 and 56 and they all had a moderate cosmetic deformity. The fifth patient had a decompensated 64 lumbar curve at the age of 14 years 7 months and was then treated by spinal fusion.

Semi-segmented hemivertebrae

Nineteen patients had a single semi-segmented hemivertebra producing a single congenital scoliosis. Seventeen of these patients were followed without treatment for a mean of 6 years 1 month (range 2 to 15 years) and two were first seen untreated at skeletal maturity.

T1 to T4. Two patients had a single semi-segmented hemivertebra, both at T3. Neither curve progressed, and at the most recent reviews at the ages of 12 and 13 years the curves measured 24 and 32 respectively.

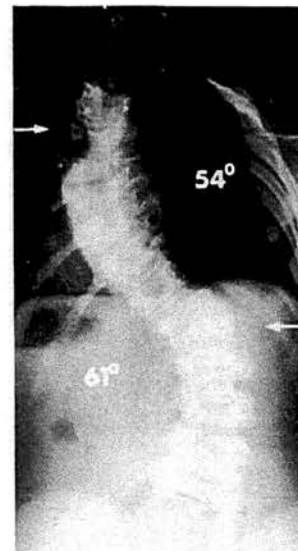


Fig. 14



Fig. 15

Case 7. Figure 14 - A girl aged 10 years with two opposing hemivertebrae occurring in different regions at T5 and L2. These hemivertebrae have produced two curves which are unbalanced. Figure 15 - The patient lists to the left and there is a significant cosmetic deformity.

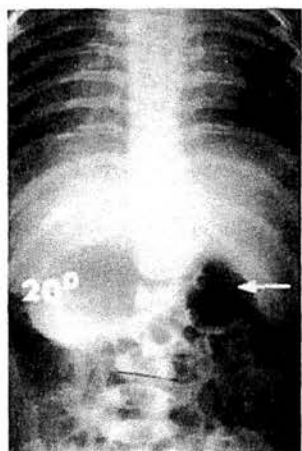


Fig. 16

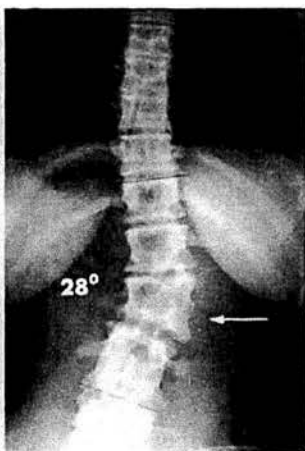


Fig. 17

Case 8. Figure 16 A girl aged 6 months with a 20° right lumbar scoliosis due to a semi-segmented hemivertebra at L2. The hemivertebra is synostosed with L3. Figure 17 No treatment was given and the curve deteriorated very slowly to measure 28° at 15 years 8 months when the patient was skeletally mature.

T5 to L1. Six patients had semi-segmented hemivertebrae in this region. Only two patients were seen before the age of 10 years when their curves were less than 25° and did not deteriorate. The other four patients were followed after the age of 10 years when their curves deteriorated at less than 1° per year. One patient was last seen at age 13 when her curve measured 27°; three patients reached skeletal maturity without treatment with a mean curvature of 34° (range 31° to 39°).

L2 to L4. Six patients had a lumbar semi-segmented hemivertebra (Figs 16 and 17). Five of them were seen before the age of 10 years; four of the curves did not progress and one deteriorated at less than 1° per year, giving a mean curvature at 10 years of age of 27° (range 18° to 36°). After the age of 10, four untreated patients were followed and the mean rate of deterioration of their curves increased to 1° per year (range 0° to 1.5°). Two patients last reviewed at 11 and 14 years had curves measuring 18° and 33° respectively, while the other two patients reached skeletal maturity without treatment with curves of 28° and 37°.

Lumbosacral. Five patients had a lumbosacral hemivertebra which was synostosed to the sacrum. These patients presented between the ages of 10 and 14 years because of the secondary structural curve above the hemivertebra and the tendency for the upper trunk to list to the side opposite the congenital anomaly. These secondary curves deteriorated at a mean rate of 2° per year (range 1.8° to 2.4°); three of the patients reached skeletal maturity without treatment with curves between 26° and 31°. The remaining two patients received treatment to control their list when they were 14 years old and their secondary structural curves measured 20° and 31°.

Incarcerated hemivertebrae

Eleven patients had a single incarcerated hemivertebra and all their curves behaved in the same manner regard-

less of their site (Figs 18 and 19). Eight patients were diagnosed in the first year of life with a mean curvature of 17° (range 11° to 22°). All 11 patients were followed without treatment; before the age of 10 eight curves did not progress and three deteriorated at less than 0.5° per year, so that at 10 years no curve exceeded 28°. Three patients were followed untreated after 10 years and their curves did not deteriorate. One patient had reached skeletal maturity with a 23° curve.

Multiple hemivertebrae

Six patients had multiple small curves due to hemivertebrae which alternated on either side of the thoracic and thoracolumbar spine. Two patients had three hemivertebrae, three had four hemivertebrae each, and one patient had six. These were a mixture of all three basic types of hemivertebrae. The resulting curves were closely associated and tended to balance each other, causing little deformity other than stunting of the spine.

DISCUSSION

This study has shown that the potential for a hemivertebra to cause a significant scoliosis depends on four factors: first, and most important, the type of hemivertebra; secondly, its site; and thirdly, the number of hemivertebrae and their relationship to each other. Finally, the age of the patient is also important because curves due to hemivertebrae often deteriorate much more rapidly during the adolescent growth spurt (Tables I, II, III and IV). It is only at skeletal maturity, when the vertebral growth plates fuse, that the potential for increasing deformity ceases.

The commonest pathological type of hemivertebra (65%) is fully segmented and non-incarcerated. Here there is absence of two growth plates on the unformed

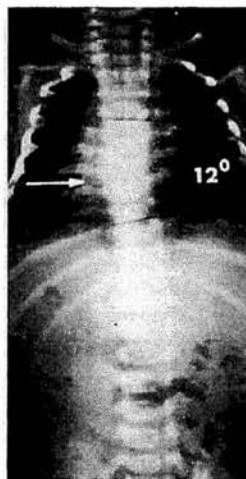


Fig. 18

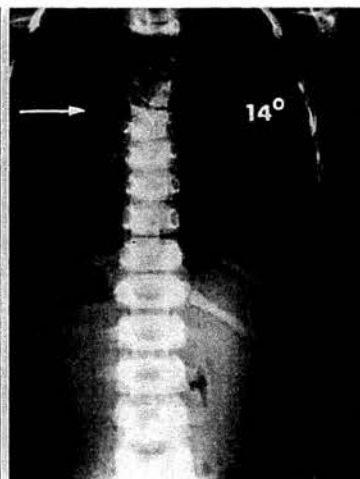


Fig. 19

Case 9. Figure 18 An infant aged two months with a 12° left thoracic scoliosis due to an incarcerated hemivertebra at T6. Figure 19 No treatment was given and there was virtually no deterioration at 13 years 8 months.

side of the hemivertebra in contrast to the two relatively normal growth plates, one on each surface of the developed part of the vertebra. This means that as the hemivertebra grows it acts as an enlarging wedge.

An incarcerated hemivertebra is the least common type (13%), and although it is fully segmented, it has a poor growth potential. The resulting scoliosis deteriorates at less than 1° per year and these curves, regardless of their site, never exceed 30° at skeletal maturity (Figs 18 and 19) so treatment is not required.

A semi-segmented hemivertebra (22%) is congenitally synostosed to one of its neighbouring vertebra and, as a result, two growth plates are absent on the convexity of the curve. This tends to balance the growth plates on the two sides of the hemivertebra although the hemivertebra itself causes tilting of the spine and can induce a progressive scoliosis (Figs 16 and 17). These curves do not exceed 40° at skeletal maturity and treatment is not usually required except occasionally when the hemivertebra is at the lumbosacral junction.

Fully segmented non-incarcerated hemivertebrae cause most problems. A single such hemivertebra is most common and the prognosis for deterioration of the resulting curve depends on the site of the hemivertebra.

Upper thoracic curves (hemivertebrae T1 to T4) deteriorate slowly but can reach 40° by skeletal maturity. Although these curves are not severe, they can cause a cosmetic deformity by elevation of the shoulder; this becomes apparent when the curve exceeds 30° and may be distressing enough in girls to warrant early prophylactic treatment. This is especially true if there is an associated Sprengel's shoulder on the same side as the hemivertebra.

Lower thoracic and thoracolumbar curves (hemivertebrae T5 to L1) deteriorate more rapidly and can exceed 45° at skeletal maturity, but these curves are not usually as rotated as an equivalent idiopathic curve and do not produce as severe a cosmetic deformity (Figs 3 and 4).

Lumbar curves (hemivertebrae L2 to L4) deteriorate relatively slowly at a rate similar to those in the upper thoracic region. They can reach 45° by skeletal maturity but cause only a relatively mild cosmetic deformity.

Lumbosacral hemivertebrae are important because they cause the lumbar spine to take off obliquely from the sacrum, resulting in the development of a large secondary structural thoracolumbar curve. This curve is usually significantly rotated and when combined with the trunk listing to the side opposite the hemivertebra, results in moderate to severe cosmetic deformity (Figs 8 and 9). These patients require early prophylactic treatment before they begin to list and before the secondary curve becomes structural.

Two unilateral fully segmented hemivertebrae are much less common but cause a much greater growth imbalance because four of the growth plates on the concavity of the curve are absent. These curves deteriorate much more rapidly than those due to a single fully segmented hemivertebra (Table II). Upper thoracic curves of this type (apex T1 to T4) usually exceed 40° by 10 years of age, causing significant cosmetic deformity due to shoulder elevation and head tilting (Figs 10 and 11). Lower thoracic and thoracolumbar curves (apex T5 to T12) deteriorate most rapidly of all at 3° per year and frequently exceed 50° by 5 years of age. Without treatment these curves could reach 70° by skeletal maturity and therefore all curves due to two unilateral fully segmented hemivertebrae require prophylactic treatment as soon as they are diagnosed.

If there are two opposing fully segmented hemivertebra the prognosis depends on whether the hemivertebrae are close together or in different regions of the spine. If they are close together they tend to balance each other and only cause two small kinks in the spine with minimal cosmetic deformity (Fig. 13). No treatment is required. If the hemivertebrae are in different regions, the resulting curves are often unbalanced and cause the trunk to list to one side (Figs 14 and 15). When this occurs prophylactic treatment is required.

Multiple hemivertebrae are often a mixture of types but the spine usually remains balanced and there is no deformity apart from some stunting of the trunk. No treatment is required.

In conclusion, it appears that incarcerated and semi-segmented hemivertebrae do not usually require treatment. The fully segmented non-incarcerated hemivertebra may require treatment, but this depends on the site, number and relationships of the anomaly or anomalies. If treatment is necessary, this should be anticipated and carried out at an early age because it is much easier to prevent than it is to correct severe deformity resulting from hemivertebrae. In planning prophylactic treatment, it should be appreciated that it is not possible to create growth on the unformed side of the hemivertebra and treatment should therefore be directed towards preventing the deforming growth of the hemivertebra.

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Congenital Scoliosis Caused by a Unilateral Failure of Vertebral Segmentation With Contralateral Hemivertebrae

Michael J. McMaster, MD, FRCS

Study Design. The medical records and serial spine radiographs of 59 consecutive patients with congenital scoliosis caused by unilateral unsegmented bar with contralateral hemivertebrae were reviewed.

Objectives. To study the presentation, natural history, and treatment of these patients.

Summary and Background Data. This is the least common type of congenital scoliosis.

Methods. The mean age at diagnosis was 4 years 1 month. Forty-three patients were observed without treatment for a mean of 6 years and 1 month. Prophylactic arthrodesis was performed in 10 patients before they were 5 years old. Thirty-five patients had a corrective procedure and arthrodesis after they were 5 years old.

Results. Thoracolumbar curves had the worst prognosis: without management all but two exceeded 50° when the patients were 2 years old. All untreated curves exceeded 88°. Midthoracic curves had only a slightly less severe prognosis, and all but one exceeded 40° by the time the patient was 2 years old. All untreated curves exceeded 70°. In eight of the 14 patients whose congenital curves had their apexes at T5, T6, or T7, a long secondary structural curve developed on the opposite side in the thoracolumbar region, and this contributed significantly to the overall deformity. Occult intraspinal anomalies were present in 24 patients (41%). Surgical treatment after the patient was 5 years old was not successful in producing significant correction of these severe rigid deformities.

Conclusion. These patients have the most rapidly progressive and severely deforming of all types of congenital scoliosis. All midthoracic, thoracolumbar, and lumbar curves require immediate prophylactic surgical treatment by anterior and posterior arthrodesis—preferably in the first year of life. [Key words: congenital scoliosis, natural history, spine arthrodesis, unilateral unsegmented bar with contralateral hemivertebrae] *Spine* 1998;23:998-1005

Congenital scoliosis caused by a unilateral unsegmented bar has a bad prognosis.^{9,16} However, there is a smaller

less well-recognized group of patients who, in addition to a unilateral unsegmented bar, have one or more hemivertebrae on the contralateral side of the spine at the same level; and they have a worse prognosis. These patients have some of the severest deformities seen by the spine surgeon.

Nasca et al¹¹ in 1975 were the first to describe congenital scoliosis caused by multiple hemivertebrae associated with a unilateral bar. They reported cases involving 18 children. In 14, the condition was diagnosed in the first 2 years of life. Adequate data were available on only 4 patients whose curves deteriorated at a mean 4° per year. When last seen, 4 patients had curves greater than 100°. In 1982, McMaster and Ohtsuka⁹ reported on the natural history of 251 patients with congenital scoliosis and noted 25 who had a unilateral unsegmented bar with contralateral hemivertebrae. They emphasized the poor prognosis for this type of scoliosis.

In this report, clinical characteristics, natural history, and management are described in 59 patients with congenital scoliosis caused by a unilateral unsegmented bar combined with contralateral hemivertebrae. Forty-two patients were observed until they reached skeletal maturity.

■ Material and Methods

The medical records and spine radiographs of 59 consecutive patients with congenital scoliosis caused by unilateral unsegmented bar with contralateral hemivertebrae at the same level, seen at the Princess Margaret Rose Orthopaedic Hospital, Edinburgh, between 1960 and 1990, were reviewed. Patients with kyphoscoliosis or myelomeningocele were excluded.

There were 45 female and 14 male patients whose mean age was 4 years 1 month (range, birth to 18 years) when spinal radiographs were available for diagnosis. Forty-three patients were observed for a mean of 6 years 1 month (range, 1 year to 13 years 3 months) before treatment. Thirteen patients, seen more recently, had immediate treatment; 3 untreated patients were first seen at skeletal maturity.

The congenital vertebral anomalies were diagnosed on the anteroposterior and lateral spine radiographs. Localized radiographs of the apex of the curve, taken obliquely with the patient supine, often provided a true anteroposterior view of the more severely rotated spine and provided a clearer picture of the vertebral anomalies. The sites of the vertebral anomalies, apical vertebra, and end vertebrae were noted. Occasionally,

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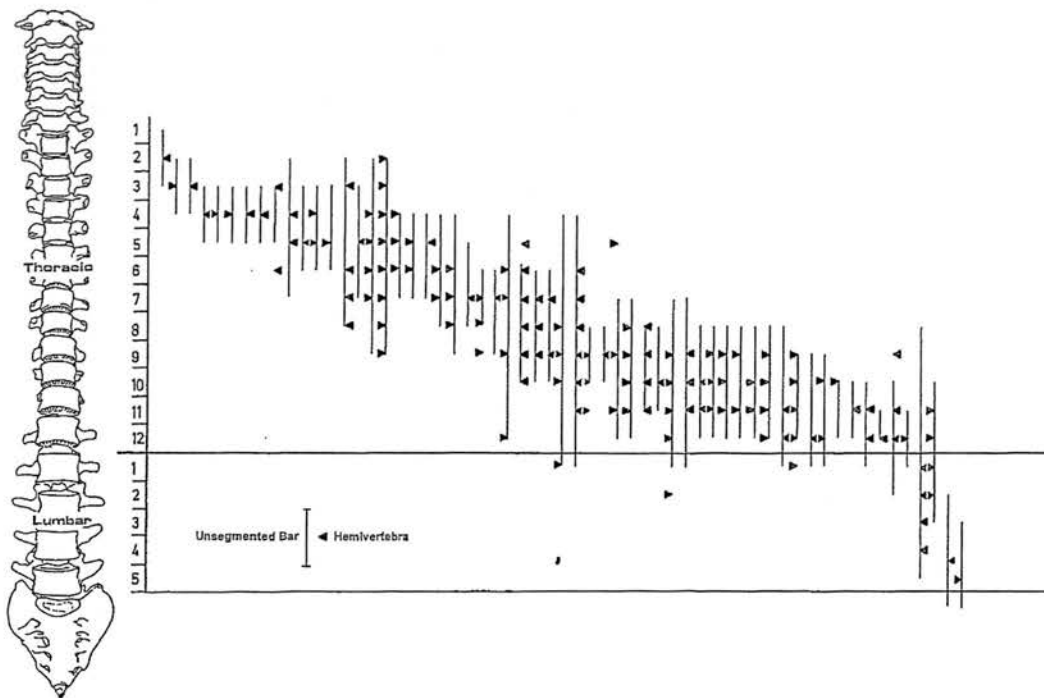


Figure 1. The extent of the unsegmented bar and the sites of the hemivertebrae in 59 patients.

additional vertebral anomalies were present at levels other than those included in the congenital curve, but these were not considered if they were not contributing to the spinal deformity. The congenital curves were grouped into four regions depending on the apex of the scoliosis: upper thoracic, apex T2 to T4; midthoracic, apex T5 to T9; thoracolumbar, T10–L1; lumbar, L2–L4, inclusive.

The Cobb angle^{2,7} of the scoliosis was remeasured on the anteroposterior radiographs of the spine taken with the patient erect at the initial visit and at the follow-up attendances, which occurred every 4–6 months. These serial spine radiographs showed the rate of deterioration of the congenital scoliosis, the development of secondary curves, any change in head tilt, shoulder asymmetry, decompensation of the trunk, and pelvic obliquity. Accurate measurement of vertebral rotation^{12,13} at the apex of the congenital curve was difficult because of the abnormal bony anatomy, which was obscured by the spinal fusion. Clinical photographs, taken when the patient was first seen, before and after surgery, and at final follow-up, were reviewed. Skeletal maturity was determined radiographically when there was complete ossification and fusion of the iliac apophysis.¹⁹

■ Results

Natural History

All 59 patients had a single congenital scoliosis. There were 30 curves to the left and 29 to the right. The extent of the unsegmented bar and the sites of the hemivertebrae for each patient are shown in Figure 1.

Upper Thoracic Scoliosis. There were nine patients in this group, and all had a unilateral unsegmented bar extending across three vertebrae. Eight of these patients had a single contralateral hemivertebra, and one patient had two contralateral hemivertebrae. Of the nine patients, only two received diagnoses before they were 10 years old. One 4-year-old patient had a 48° scoliosis that was managed immediately by arthrodesis. The second patient, aged 5 years 10 months, had a 45° scoliosis which deteriorated at 1.5° per year, and when the patient was 10, measured 51°. After the patient was 10, the rate of deterioration increased to 8° per year; arthrodesis was performed when he was 13 years 10 months old and the curve was 83°.

The remaining seven patients did not receive diagnoses until after they were 10 years old. Two of these patients were treated by arthrodesis when first seen at age 13 years with curves of 70° and 80°, respectively. Five patients were not treated and were observed to skeletal maturity, when their curves measured 40°, 42°, 42°, 46°, and 50°, respectively.

An elevation of the shoulder line on the convex side of the scoliosis was present in all of these patients and produced a significant deformity. Tilting of the head away from the convexity of the curve occurred in three patients.

Midthoracic Scoliosis. There were 28 patients in this group. They had a more extensive unilateral unseg-

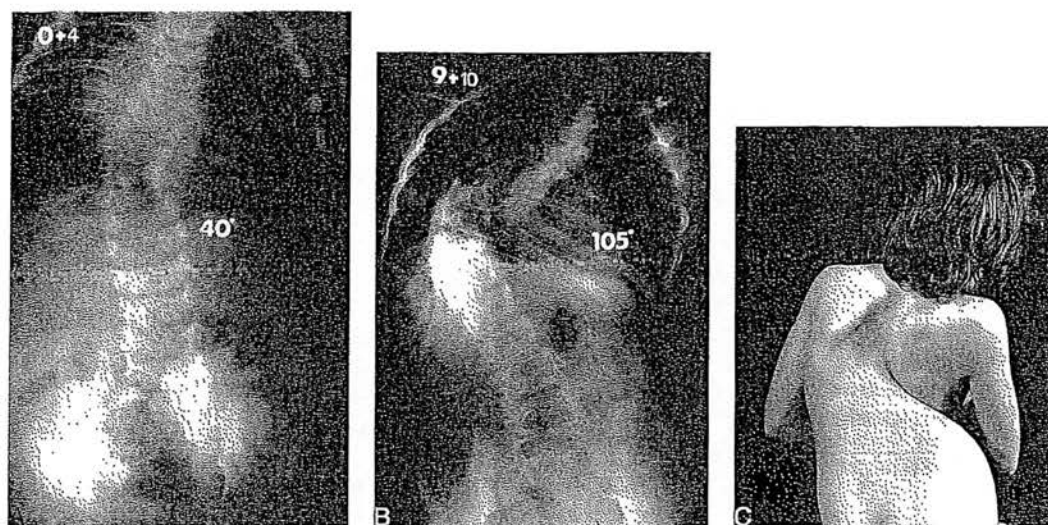


Figure 2. A, An infant aged 4 months with a 40° left thoracic scoliosis caused by a unilateral unsegmented bar with three contralateral hemivertebrae. Spina bifida occulta extends throughout the lumbar region, but no intraspinal anomaly is present. No treatment was given. B, When the patient was aged 9 years 10 months, the scoliosis had increased to 105°. C, There is a severe cosmetic deformity caused by shoulder imbalance, distortion of the rib cage, and malalignment of the trunk.

mented bar extending across a mean of five vertebrae (range, 3–10 vertebrae) with 2.8 contralateral hemivertebrae (range, 1–6 hemivertebrae).

Twenty-one patients were observed without treatment before they were 10 years old (Figures 2A–C) for a mean of 5 years 6 months (range, 1–10 years), during which time the median rate of deterioration of their congenital scoliosis was 5° per year (range, 2–10° per year). Fourteen of these patients were diagnosed in the first year of life with a mean scoliosis of 48° (range, 23–100°). There were 19 patients, untreated at age 2 years, whose mean curve was 56° (range, 37–100°). Ten patients were treated by arthrodesis before they were 8 years old (mean age, 3 years 9 months), when their mean curve was 73° (range, 42–120°). An additional 7 patients, between 8 and 10 years old, had arthrodesis when their mean curve was 71° (range, 53–99°). There were 5 untreated 10-year-old patients whose mean curve was 64° (range, 60–70°).

Five patients older than 10 years were observed untreated for a mean 3 years 9 months (range, 2 years 8 months to 7 years) and the median rate of deterioration of their scoliosis increased to 8° per year (range, 5–10° per year). Five patients had arthrodesis at a mean age of 12 years 10 months (range, 11 years 8 months to 13 years 6 months), when their mean curve was 78° (range, 70–88°). Three patients were untreated at skeletal maturity with curves of 75°, 83° and 132°.

An elevation of the shoulder line on the convexity of the curve occurred in all of these patients, and 17 had significant trunk decompensation (more than 3 cm).

Secondary structural thoracolumbar scoliosis developed below the congenital scoliosis on the opposite side of the spine in eight of these patients during the first few

years of life (Figures 3A and 3B). This secondary curve extended across a mean of seven vertebrae (range, six to seven vertebrae) and was present in three of the six patients whose congenital curve apex was at T5, in all four patients whose congenital curve apex was at T6, and in one of the four patients with curve apex at T7. Why such a curve should develop in some of these patients and not in others is unknown. The secondary curve did not contain any congenital vertebral anomalies and was initially compensatory and correctable. However, it later rotated and became fixed, producing a rib hump and a significant additional deformity. In three patients, the secondary structural curve became larger than the congenital scoliosis before they were 5 years old.

Thoracolumbar Scoliosis. There were 20 patients in this group. The unilateral unsegmented bar extended across a mean five vertebrae (range, 2–10 vertebrae) with a mean 2.5 contralateral hemivertebrae (range, one–five hemivertebrae).

Sixteen patients were observed without treatment before they were 10 years old for a mean of 5 years (range, 1–9 years), during which time the median rate of deterioration of their scoliosis was 7° per year (range, 3–13° per year). Eleven patients in the first year of life had a mean curve of 45° (range, 22–60°). Sixteen untreated patients had a mean curve of 58° (range, 42–76°) by age 2 years. Between the ages of 2 and 8 years, four of these patients had arthrodesis at a mean age of 5 years 5 months when their mean curve was 100° (range, 74–142°). Seven patients had arthrodesis between the ages of 9 and 10 years, when their mean curve was 85° (range, 66–107°).

Three patients older than 10 years were observed

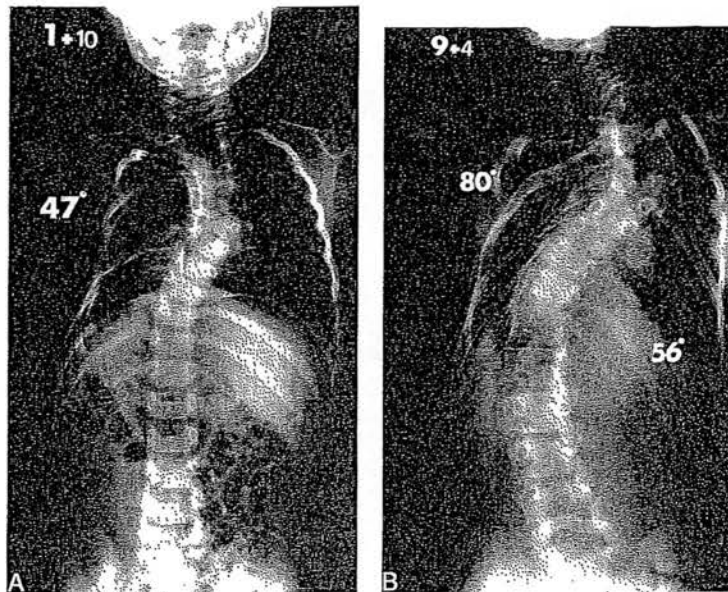


Figure 3. A, An infant aged 1 year 10 months with a 47° right thoracic scoliosis (apex T5) caused by a unilateral unsegmented bar with two contralateral hemivertebrae. No treatment was given. B, When the patient was aged 9 years 4 months, the congenital scoliosis had increased to 80° and a secondary structural thoracolumbar curve had developed on the opposite side. The patient is severely deformed.

without treatment for between 3 years 8 months and 4 years 5 months when the median rate of deterioration of their curves increased to 14° per year (range, 10–15°). Six patients had arthrodesis between the ages of 12 and 16 years, when their mean curve was 115° (range, 88–130°).

All of the patients had a significant cosmetic deformity caused by the severity of their scoliosis combined with shoulder imbalance, distortion of the rib cage and decompensation of the trunk. Pelvic obliquity was present in 12 patients. Right patients with pelvic obliquity also had a true leg length discrepancy. In the remaining 4 patients, the pelvic obliquity produced an apparent leg length discrepancy with no true shortening.

Lumbar Scoliosis. There were only two patients in this group. One patient had a 16° scoliosis at birth caused by an unsegmented bar extending from L3 to the sacrum and a single contralateral hemivertebra at L5. This curve deteriorated rapidly to 77° at age 5 years, when she had a very severe deformity caused by listing of the trunk and pelvic obliquity producing an apparent leg length discrepancy.

The second patient, 2 years old, had 21° scoliosis caused by an unsegmented bar extending from L2 to the sacrum, with a single contralateral hemivertebra at L4. She was treated by arthrodesis at age 3 years 10 months when the scoliosis was 33° and accompanied by pelvic obliquity and listing of the trunk.

Intraspinal Anomalies. Twenty-four (41%) of the 59 patients had a congenital intraspinal anomaly diagnosed by a myelogram or magnetic resonance scan. These anomalies occurred most frequently in association with thoracolumbar curves (11 patients).

Diastematomyelia was seen in 23 of the 24 patients.

The spur occurred at all levels from T6 to L5, but was most frequent between T11 and L3, inclusive (18 patients; 78%). In 12 patients, the diastematomyelia spur was at the same level as the vertebral anomalies causing the scoliosis, and in the remaining 11 it was at a lower level. Spina bifida occulta was present in 22 of these patients, and the laminar defect affected from 2 to 10 adjacent vertebrae.

Three patients had an additional intraspinal anomaly occurring at the same level as the diastematomyelia; one patient had an intradural lipoma and tethered cord, another had an epidermoid cyst, and the third had neural adhesions. A fourth patient who had diastematomyelia at T12 also had a neurenteric cyst at T3–T4. The 24th patient, who did not have diastematomyelia, had a tethered cord with unilateral conjoined nerve roots in the lumbar region.

Cutaneous Abnormalities. Nineteen (79%) of the 24 patients with intraspinal anomalies had an abnormality of the skin or subcutaneous tissues overlying the spine. This cutaneous abnormality was in the same region as the intraspinal anomaly in 14 patients (58%) and at a different level in 5 patients. The most common cutaneous abnormalities were a hairy patch (10 patients), red nevus (7 patients) and, less frequently, a fat pad (one patient) or a dimple (one patient). It should be noted that 6 other patients without myelographic evidence of an intraspinal anomaly also had a cutaneous abnormality overlying the spine.

Neurologic Abnormalities. Twelve (50%) of the 24 patients with intraspinal anomalies had neurologic abnormalities affecting one leg or the bladder. The neural deficit was mild in five patients who had unilateral reflex

abnormalities, mild hypoplasia of the leg and foot combined with a mild pes cavus and claw toes; moderate in four patients who had more severe muscle weakness below the knee, resulting in an equinovarus deformity and hypoaesthesia of the foot; severe in three patients who had paresis of the lower leg and urinary incontinence.

Neurologic deterioration occurred in only two patients. One patient with unilateral lower limb weakness developed urinary incontinence at 4 years of age and the second patient with a paralytic foot deformity was thought to have increasing weakness at age 2 years. An additional five patients, whose myelograms showed no evidence of an intraspinal anomaly, also had mild unilateral lower limb neurologic abnormalities.

Rib, Chest, and Shoulder Anomalies. Congenital rib fusions were present in 34 patients (58%) and occurred on the concavity of the scoliosis in all but 2 patients. Spreading of the ribs or an absence of several adjacent ribs was present in 8 patients and was always on the concave side of the scoliosis (Figure 3A).

A Sprengel's shoulder deformity was present in six patients, all of whom had thoracic scoliosis. The congenital elevation of the scapula was on the concavity of the scoliosis in five patients and partly balanced the elevated shoulder line caused by the curvature. In the sixth patient, the elevated scapula was on the convex side of the curve and increased the deformity.

Other Congenital Anomalies. Heart and renal anomalies were diagnosed in three patients each. Congenital dysplasia of the hip was present in three patients and a coxa valga deformity in one. Hypoplasia of the arm occurred in one patient and hypoplasia of the thumb in one patient. A hiatal hernia was present in three patients. Other anomalies, each affecting one patient were, a cleft palate, diaphragmatic hernia, and congenital hepatic fibrosis.

Surgical Treatment

Excision of Intraspinous Anomaly. Fifteen patients had an excision of a diastematomyelia spur or other intraspinal anomaly. These operations were performed for one of three reasons. The first was because of a progressive neurologic deficit that occurred in two patients less than 5 years old; the operation was successful in preventing further neurologic deterioration, but there was no improvement. The second was as a prophylactic measure to prevent possible neurologic deterioration in five patients with stable neurology whose intraspinal anomalies were diagnosed before they were 2 years old; two of these patients had mild unilateral lower limb neurologic abnormalities, and the other three were normal. The intraspinal anomaly was excised at a mean age of 10 months (range, 2 months to 1 year 3 months), and after surgery there was no change in their neural status. The third and most common reason was to prevent possible neural complications occurring during curve correction and arthrodesis in eight patients; the mean age at the time of the neurosurgical procedure was 6 years 8 months (range, 2 years 8 months to 10 years 10 months), and after surgery there was no neurologic deterioration in any of the patients.

Prophylactic Early Spine Arthrodesis. Ten patients less than 5 years old had arthrodesis in an attempt to prevent further deterioration of their deformities. No instrumentation was used; after surgery, an underarm plaster jacket was worn for 6 months. Intraspinous anomalies were diagnosed in six of these patients, but excised in only four before their arthrodesis.

A posterior arthrodesis alone was performed in five patients at a mean age of 2 years 2 months (range, 1 year 3 months to 4 years 3 months), when the mean curvature was 47° (range, 33–68°). The spine was arthrodesed throughout the length of the congenital curve, from the upper- to the lower-end vertebrae. Allograft bank bone from degenerate femoral heads was used as graft material. The length of the fusion was across a mean of eight vertebrae (range, 6–10 vertebrae) and extended from one to two levels above to one to three levels below the unsegmented bar. One patient with a Sprengel deformity on the convexity of the curve had her elevated scapula reduced by a Woodward procedure¹ performed at the same time as the spine surgery.

After surgery, the patients were observed for a mean 6 years 4 months (range, 3 years to 12 years 6 months), and a solid fusion developed in all. In none of these patients were additional vertebrae added to the curve. The mean loss of correction in the fused segment of the spine was 5° (range, 3–9°). There was no apparent increase in spine rotation, although measurement of vertebral rotation was difficult because the bony anatomy became obscured by the fusion. A combined anterior and posterior arthrodesis was performed in five patients under the same anesthetic at a mean age of 2 years 4 months (range, 7 months to 4 years 8 months), when their mean curve was 67° (range, 44–108°). The rib excised during the first stage thoracotomy was used as graft material supplemented by allograft bank bone as necessary.

In two patients, the arthrodesis extended from one level above to one level below the unilateral unsegmented bar, but this was insufficient to stabilize the deformity. Both patients required additional surgery after 1 and 5 years to extend the fusion to include additional vertebrae above and below.

The remaining three patients had a more extensive anterior and posterior arthrodesis. The length of the fusion was across a mean nine vertebrae (range, 6–12 vertebrae) and extended over the whole length of the measured curve from the upper- to the lower-end vertebrae, which were two to three levels above and below the unsegmented bar. These patients have a stable deformity and have been observed for a mean of 4 years 9 months (range, 3–9 years).

Posterior Arthrodesis and Correction in a Localizer Jacket. Eighteen patients had posterior arthrodesis followed by correction in a Risser-type localizer jacket at a mean age of 10 years 3 months (range, 6 years to 13 years 10 months), when their mean curve was 72° (range, 46–90°). Intraspinous anomalies were diagnosed in eight of these patients but were excised in only three before the arthrodesis. Autologous bone from the iliac crest was used as bone graft material, and the spine was arthro-

desed from the upper to the lower neutral vertebrae. After surgery, a Risser localizer jacket was applied with the patient under light sedation on a traction table, and the scoliosis was reduced by a mean 10° (range, 0–26°). There was no change in the neural status of any of these patients; three had a preoperative neural deficit, and 15 were normal.

The Risser jacket was removed after 9 months. Follow-up observation was done for a mean 6 years 9 months (range, 2 years 6 months to 10 years 6 months), and a solid fusion developed in all patients. The mean loss in correction at final follow-up was 14° (range, 10–23°) with the greatest loss occurring in the first year. No patient required an extension of the spinal fusion. All of the patients were observed to skeletal maturity, and all remained severely deformed.

Posterior Arthrodesis With Harrington Instrumentation. Eight patients had a posterior arthrodesis with intraoperative correction, using Harrington instrumentation at a mean age of 11 years 7 months (range, 6 years 2 months to 15 years), when the mean curve was 87° (range, 56–130°). Three of these patients had a diastematomyelia spur removed before their arthrodesis. Autologous bone from the iliac crest was used as graft material. The spine was arthrodesed and instrumented from the upper to the lower neutral vertebrae. The mean curve was reduced to 67° (range, 40–90°). An intraoperative wake-up test¹⁵ was performed, and there was no change in their neural status; before surgery five patients had a neural deficit and three were normal. Intraoperative spinal cord monitoring using somatosensory-evoked potentials has been used since 1980.

After surgery, patients wore an underarm plaster jacket for 6 months. Follow-up observation was done for a mean 5 years 6 months (range, 3–8 years), and a solid fusion developed in all. The mean loss in correction at final follow-up examination was 7° (range, 1–15°). All had reached skeletal maturity, and all were grossly deformed.

Combined Anterior and Posterior Arthrodesis With Harrington Instrumentation. Three patients had anterior arthrodesis, followed 5 days later by posterior arthrodesis with Harrington instrumentation. The mean age of the patients at surgery was 10 years (range, 6 years 5 months to 14 years 5 months), when the mean curve was 102° (range, 98–107°). Harrington instrumentation was applied to act as an internal strut, followed by an intraoperative wake-up test¹⁵ to check neurologic status. Spinal cord monitoring by somatosensory-evoked potentials also was performed throughout the procedure. After surgery, the mean curve was reduced to 76° (range, 74–80°), and neural status remained unchanged; one patient had a preoperative neural deficit, and two were normal.

An underarm plaster jacket was worn for 6 months. Follow-up observation was done for a mean 6 years 3 months (range, 4–12 years), and all had a solid fusion. The mean loss in correction was 4° (range, 6–8°), and all reached skeletal maturity. All of the patients were severely deformed.

Spine Osteotomy and Arthrodesis. Six patients with very severe deformities had a closing-wedge spinal osteotomy

performed by anterior and posterior procedures separated by 1 week. Posterior arthrodesis was also performed from the upper to the lower neutral vertebra at the time of the second procedure. Three of these patients had a diastematomyelia spur excised before the osteotomy. Compression instrumentation was used to close the osteotomy at the time of the posterior fusion; all of the patients had spinal cord monitoring and an intraoperative wake-up test. The mean age at osteotomy was 8 years 4 months (range, 4 years 1 month to 13 years 10 months), when the mean curve was 111° (range, 90–142°). After surgery, the mean scoliosis was reduced to 70° (range, 28–105°). Patients wore an underarm plaster jacket for 6 months.

One patient who had been neurologically normal had partial paralysis in one leg after the anterior osteotomy at T11–T12. No intraoperative correction had been attempted at this stage. This complication was thought to be caused by impairment of the blood supply to one half of a bifid spinal cord at the level of vertebral resection. There was no intraspinal spur. There was no further neural deterioration after the second-stage posterior osteotomy and correction of the deformity, which was carried out 3 weeks later. Sensation rapidly returned to normal, and the motor power in the leg returned to Grade IV in all muscle groups after 1 year; however, there was generalized spasticity. In the remaining five patients there was no neural change after the osteotomy; four patients had a preoperative neural deficit, and one was normal.

Follow-up observation was done for a mean 6 years 4 months (range, 2 years 6 months to ten years), during which time there was a 9° (range, 5–18°) loss of correction. Five patients were seen at skeletal maturity, and all were severely deformed.

Treatment of the Secondary Structural Thoracolumbar Scoliosis. Four patients who had arthrodesis of their congenital scoliosis also had secondary structural thoracolumbar scoliosis, which was not included in the fusion. The mean age at surgery was 8 years 8 months (range, 4 years 8 months to 12 years). After surgery, a localizer jacket was applied without internal fixation. Before surgery, the mean size of the congenital scoliosis was 81° (range, 60–108°), and the secondary thoracolumbar curve was 54° (range, 50–65°). After surgery, the patients were observed for a mean 5 years 10 months (range, 3 years to 7 years 7 months), at which time the mean congenital scoliosis was 72° (range, 62–78°). Fusion of the congenital curve did not control the unfused secondary thoracolumbar scoliosis, which continued to deteriorate once the plaster jacket was removed and measured 65° (range, 58–74°) at final follow-up examination.

■ Discussion

This report describes 59 patients with congenital scoliosis caused by a unilateral failure of vertebral segmentation, resulting in a bar of bone joining together several adjacent vertebrae on one side of the spine, combined with one or more hemivertebrae on the opposite side at the same level (Figure 2A). The unilateral unsegmented bar does not contain epiphyseal plates and therefore does not grow longitudinally, whereas the hemivertebrae produce a greater degree of growth on the contralateral side of the spine than if there had been an unsegmented bar alone. The crankshaft effect produced by the anterolateral growth of the hemivertebrae, combined with the tethering effect of the unseg-

mented bar on the concavity, results in the most rapidly progressive and severely deforming of all types of congenital scoliosis (Figures 2B and 2C).

Radiographs of 18 infants taken shortly after birth showed that they already had a mean scoliosis of 46°. Unfortunately, the significance of the congenital vertebral anomalies was not always appreciated, and many children were not referred until the clinical deformity had become obvious. A few initially were misdiagnosed as having hemivertebrae alone, because the unsegmented bar did not become apparent until it was more fully ossified. In their later childhood, the radiologic diagnosis became more difficult as the spine rotated and the hemivertebrae became increasingly obscured (Figure 2B).

A thorough knowledge of the natural history is essential in planning treatment. Forty-three children were observed without treatment for a mean of 6 years 1 month. Many of these untreated patients were seen at a time when the prognosis for this type of congenital scoliosis was not fully appreciated. Thoracolumbar curves had the worst prognosis, and curves in all but two patients exceeded 50° by the age of 2 years. Their median rate of deterioration was 7° per year before they were 10 years old, increasing to 14° per year during the adolescent growth spurt, after which all untreated curves exceeded 88°. Midthoracic curves had only a very slightly less severe prognosis; all but one exceeded 40° when the patient reached age 2. The median rate of deterioration was 5° per year before the patient was 10 years old and 8° thereafter; all untreated curves exceeded 70°. Patients with midthoracic, thoracolumbar, and lumbar curves all became severely deformed at an early age, because of a combination of shoulder imbalance, severe distortion of the rib cage, decompensation of the trunk, and often, pelvic obliquity that produced an apparent leg length discrepancy (Figure 2C). Upper thoracic curves usually appeared later, with an elevation of the shoulder line on the convexity occasionally accompanied by tilting of the head.

An additional spine deformity occurred in 8 of the 14 patients whose congenital scoliosis had its apex at T5, T6, or T7. A long secondary curve developed on the opposite side in the thoracolumbar region. This curve, which did not contain congenital anomalies, was initially compensatory and correctable. However, it deteriorated rapidly, becoming rotated and fixed, and contributed significantly to the overall deformity (Figures 3A and 3B). It is possible that this secondary scoliosis occurred because of transmission of the crankshaft effect produced by the midthoracic congenital vertebral anomalies to the more mobile thoracolumbar region.

The development of the spinal cord is closely associated with that of the vertebral column, and 24 patients (41%) in this series also had an intraspinal anomaly with diastematomyelia being most frequent. Myelographic examination was used in most of these patients, but the more modern technique of magnetic resonance imaging is much superior and may have detected an even higher incidence.

It is accepted that an intraspinal anomaly, such as a diastematomyelia spur that tethers the neural structures, should be excised if there is a progressive neurologic deficit or before attempting to correct an associated spi-

nal deformity.¹⁰ There is also neurosurgical opinion³ that suggests that the mere presence of a potentially tethering intraspinal lesion is sufficient reason for prophylactic surgical treatment, which is best performed as early as possible and before there is neural dysfunction. In this series, the intraspinal anomaly was excised from five children with stable neurology before the age of 2 years as a prophylactic measure and from two children with deteriorating neurology before the age of 5 years. There was no neurologic deterioration after these procedures, and the neurology stabilized in the two children who had a progressive deficit. All of the patients who had intraoperative spine instrumentation to correct their deformity had their intraspinal anomalies excised before arthrodesis. There were no neurologic complications. Excision of the anomaly combined with spinal cord monitoring using somatosensory-evoked potentials or an intraoperative wake-up test¹⁵ is recommended to prevent traction on the neural structures from occurring during corrective surgery.⁸

In planning treatment, it is important to recognize that from an early age, a unilateral unsegmented bar with contralateral hemivertebrae is an extensive rigid deformity. Once significant scoliosis has developed, it can only be corrected by a spinal osteotomy, which can be a difficult and dangerous procedure. It is not possible to create longitudinal growth within the unsegmented bar, because it does not contain epiphyseal plates. The only way to prevent deterioration is to stop further growth on the convex side opposite the bar, and this requires a solid, stable fusion. Bracing is totally ineffective, and surgery should be performed as early as possible. There is no perfect treatment, and the best that can be achieved is a short, relatively straight spine that is balanced.¹⁷

Prophylactic fusion of the congenital scoliosis was performed before the age of 5 years in 10 children whose mean curve was 57°. Five had a posterior arthrodesis alone and 5 had a combined anterior and posterior arthrodesis. At best, an early fusion will only maintain the status quo. Unfortunately, many of these children were already too deformed for the procedure to be ideal. The congenital scoliosis contains not only the anomalous segment but also of a number of relatively normal vertebrae at the upper and lower ends, which are also tilted into the curve. Failure to include all of these vertebrae in the fusion resulted in continued progression of the scoliosis in 2 children who required an extension of their fusions. In this type of congenital scoliosis, the surgeon is unlikely to regret operating too early or fusing too many vertebrae, but will always regret operating too late or having fused too few vertebrae. Sprengel's deformity of the scapula on the convex side of the scoliosis, which is contributing to the deformity, should also be surgically reduced at the time of the posterior arthrodesis.¹

Loss of correction after an early posterior spine fusion has been attributed to a weak fusion mass¹⁷ or to rotation of the spine as a consequence of the crankshaft effect produced by anterior spine growth in the presence of a posterior tether caused by the fusion.^{4,6} Combined anterior and posterior arthrodesis has the advantage of directly overcoming a possible crankshaft effect and pro-

ducing a more solid fusion. The rib excised at the time of the thoracotomy provides a good source of autologous bone graft, which may not be available from the iliac crest in these very young children. However, a posterior fusion alone can produce an acceptable result, providing there is a very strong, thick fusion capable of overcoming the anterior growth of the spine. If the curve continues to deteriorate, early exploration and reinforcement of the posterior fusion is essential to detect pseudarthrosis or to strengthen the fusion by the addition of further graft material. Five children in this series had early posterior fusion alone, and none of their curves progressed more than 8° or showed signs of increasing rotation. Similar results were obtained by Terek et al¹⁴ who studied the crankshaft phenomenon after early fusion in congenital scoliosis and found that only one of nine curves caused by a unilateral bar with contralateral hemivertebrae progressed more than 10° or showed signs of increasing vertebral rotation after a posterior fusion before the patient was 10 years old.

The secondary structural thoracolumbar curve, which can develop below a congenital scoliosis with its apex at T5, T6, or T7, also requires prophylactic treatment. This secondary deformity is not controlled by early fusion of the primary congenital scoliosis. Bracing has no role to play in the treatment of the congenital curve, but it can be of value in controlling the secondary curve and can postpone extending the fusion to the lower lumbar region, especially if many years of further growth are anticipated. Alternatively, it is possible at the time of posterior arthrodesis of the congenital curve, to apply pediatric spine instrumentation across both curves without fusing the secondary curve. Subsequent serial extension of the instrumentation would allow further longitudinal growth in the unfused thoracolumbar curve before extending the fusion. This technique has been applied to more recent patients who have not been included because of too short a follow-up period.

Unfortunately, most patients in this series did not receive appropriate early prophylactic treatment, and spine surgery after they were 5 years old^{5,18} was not successful in producing significant correction of these very severe rigid deformities.

Conclusion

A unilateral unsegmented bar with contralateral hemivertebrae has the worst prognosis for any type of congenital scoliosis. If left untreated, all of these curves will deteriorate rapidly to become an extremely severe rigid deformity early in the patients' life. At this stage the scoliosis can only be managed by potentially dangerous, complex salvage surgery with moderate improvement. The key to optimal treatment is early diagnosis, an appreciation of the high incidence of occult intraspinal anomalies, and an immediate anterior and posterior arthrodesis to balance the growth of the spine, preferably in the first year of life for all midthoracic, thoracolumbar, and lumbar curves.

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Natural History of Congenital Kyphosis and Kyphoscoliosis

A STUDY OF ONE HUNDRED AND TWELVE PATIENTS*

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Abstract

Background: Congenital kyphosis and kyphoscoliosis are much less common than congenital scoliosis. However, they are potentially more serious because compression of the spinal cord and paraplegia sometimes develop. The goals of the present study were to document the natural history of congenital kyphosis and kyphoscoliosis and to determine the stage at which the natural progression should be interrupted by treatment.

Methods: We reviewed the medical records and radiographs of the spine of 112 consecutive patients. Sixty-eight patients had a type-I kyphosis due to an anterior failure of vertebral-body formation, twenty-four had a type-II kyphosis due to an anterior failure of vertebral-body segmentation, and twelve had a type-III kyphosis due to a combination of anomalies; the deformities of the remaining eight patients could not be classified.

Eighty-five skeletally immature, untreated patients were first evaluated at a mean age of six years and nine months (range, two months to sixteen years and three months), and twenty-seven patients were skeletally mature at the time of the first visit. Sixty-three of the eighty-five skeletally immature patients were observed without any treatment for a mean period of six years and six months (range, one to sixteen years) before skeletal maturity, and the remaining twenty-two patients had a posterior arthrodesis of the spine soon after the initial visit. At skeletal maturity, forty-one patients had not been treated and sixty-eight had had an arthrodesis of the spine. The remaining three patients had not yet reached skeletal maturity at the time of the most recent follow-up.

Results: The apex of the kyphosis was seen at all levels but was most frequent between the tenth thoracic and the first lumbar level (seventy-four patients; 66 percent). There was no relationship between the severity of the kyphosis and its location in the spine. Progression of the curve was most rapid during the adolescent growth spurt and stopped only at skeletal

maturity. Progression was most rapid and the magnitude of the curve was the greatest in type-III kyphosis (twelve patients) followed by type-I kyphosis due to a posterolateral quadrant vertebra (thirty-nine patients), a posterior hemivertebra (eight patients), a butterfly vertebra (fifteen patients), and a wedged vertebra (six patients). A kyphosis due to two adjacent type-I vertebral anomalies progressed more rapidly and produced a more severe deformity than did a similar single anomaly. The prognosis for type-II kyphosis was variable and was much more severe when an anterolateral unsegmented bar had produced a kyphoscoliosis (nine patients) than it was when a midline anterior bar had produced a pure kyphosis (fifteen patients), which usually progressed slowly. Spontaneous neurological deterioration due to compression of the spinal cord occurred in ten patients (seven of whom had a type-I kyphosis and three of whom had an unclassifiable anomaly) at a mean age of thirteen years and eight months, and one other patient (with an unclassifiable anomaly) had spastic paraparesis at the age of twenty-eight years.

Conclusions: Congenital kyphosis and kyphoscoliosis are uncommon deformities with the potential to progress rapidly, resulting in severe deformity and possible neurological deficits. A thorough knowledge of the natural history is essential in the planning of appropriate and timely treatment to prevent progression of the deformity and neurological complications.

A congenital kyphosis or kyphoscoliosis is an uncommon deformity in which there is an abnormal posterior convex angulation of a segment of the spine due to developmental vertebral anomalies that impair longitudinal growth anterior or anterolateral to the transverse axis of vertebral rotation in the sagittal plane. These vertebral anomalies are present at birth, hence the term congenital, but the deformity may not become clinically apparent until later in childhood.

Congenital kyphosis and kyphoscoliosis are much less common than congenital scoliosis²², but they are potentially more serious because they sometimes lead to compression of the spinal cord and paraplegia²⁸. The first description of congenital kyphosis was by Von Rokitsansky²⁰ in 1844; however, it was not until 1932 that Van Schrick¹⁹ differentiated a failure of vertebral-body formation from a failure of vertebral-body segmentation as a cause of congenital kyphosis. He reported on

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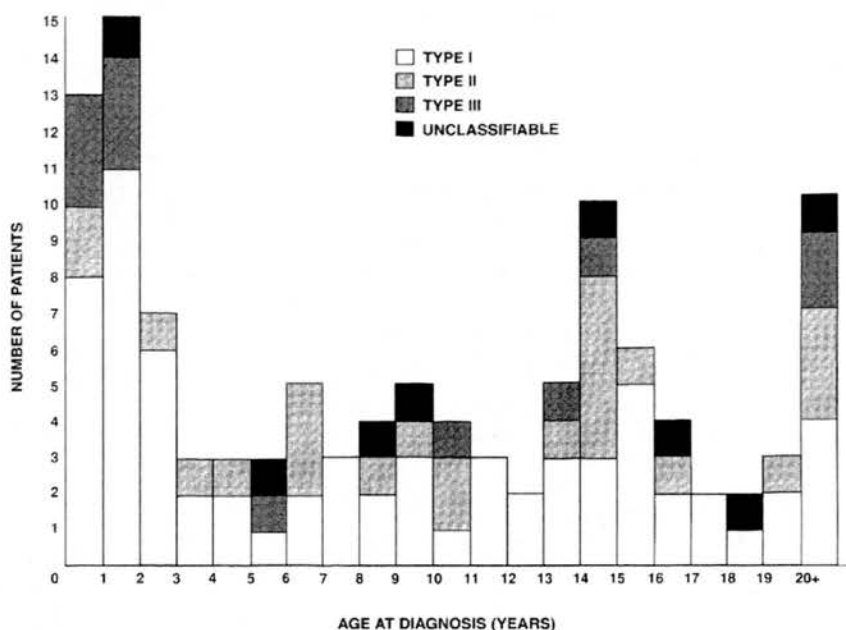


FIG. 1

Histogram showing the number of patients who had each type of congenital kyphosis and kyphoscoliosis and the ages of the patients at the time of diagnosis.

four patients and reviewed the literature, in which he identified the cases of fourteen patients. In 1955, James⁷ described twenty-one patients who had a congenital kyphoscoliosis; five became paraplegic. To our knowledge, the only study on the natural history of congenital kyphosis and kyphoscoliosis in a large series was that by Winter et al.²³, published in 1973. Those authors reported on 130 patients from several centers, but only thirty of the patients were followed without treatment for more than one year.

The goal of the present study was to further evaluate the natural history of congenital kyphosis and kyphoscoliosis. It should be kept in mind that once a patient begins treatment (which may consist only of follow-up evaluations and observation) the course of the deformity is, by definition, no longer the natural history.

Materials and Methods

We reviewed the medical records and radiographs of the spine of 112 consecutive patients who had a congenital kyphosis or kyphoscoliosis and were seen at the Princess Margaret Rose Orthopaedic Hospital in Edinburgh between 1960 and 1996. These patients were part of a group of 584 consecutive patients with congenital deformities of the spine who were studied by the senior one of us (M. J. McM.); 472 of those patients had a pure scoliosis^{9,12}, seventy-six had a kyphoscoliosis, and thirty-six had a pure kyphosis. Care was taken to exclude patients who had a scoliosis and a marked vertebral rotation that could mimic a kyphosis in the sagit-

tal plane. Patients who had a kyphosis associated with a myelomeningocele, Scheuermann disease, neurofibromatosis, skeletal dysplasia, infection, or trauma were also excluded.

Our present policy is to manage patients who have a congenital kyphosis or kyphoscoliosis at an early stage, preferably when they are less than five years old, with a posterior arthrodesis of the spine, before the kyphosis exceeds 45 degrees²⁵. A kyphosis that does not reduce to less than 45 degrees, as measured on a lateral radiograph of the spine made with the patient supine, necessitates an anterior release and an arthrodesis with strut-grafting followed by a posterior arthrodesis, often with instrumentation, to correct and stabilize the deformity. Many patients in our series did not receive optimum treatment because they were referred from other centers, where they had been followed with serial radiographs of the spine but had not received treatment.

There were sixty-four female and forty-eight male patients. Twenty-seven untreated patients were first evaluated after skeletal maturity; six of these patients subsequently had anterior and posterior procedures on the spine to correct the deformity. The remaining eighty-five untreated patients were skeletally immature when they were first evaluated, at a mean age of six years and nine months (range, two months to sixteen years and three months) (Fig. 1). Twenty-two of these skeletally immature patients had an arthrodesis of the spine soon after the initial visit. A posterior procedure was performed to prevent further progression in nine patients

TABLE I
NUMBER OF PATIENTS WHO HAD EACH TYPE OF
CONGENITAL KYPHOSIS AND KYPHOSCOLIOSIS

Type of Deformity	No. of Patients
Type I: Anterior failure of vertebral-body formation	68
Posterolateral quadrant vertebra	
Single vertebra	34
Two adjacent vertebrae	5
Posterior hemivertebra	
Single vertebra	4
Two adjacent vertebrae	4
Butterfly (sagittal cleft) vertebra	15
Anterior or anterolateral wedged vertebra	
Single vertebra	2
Two adjacent vertebrae	4
Type II: Anterior failure of vertebral-body segmentation	24
Anterior unsegmented bar	15
Anterolateral unsegmented bar	9
Type III: Mixed anomalies (anterolateral unsegmented bar with contralateral posterolateral quadrant vertebrae)	12
Type IV: Unclassifiable anomalies	8
Total	112

who had a kyphosis of less than 45 degrees, and thirteen patients with a more severe kyphosis had more complex anterior and posterior procedures on the spine to correct the deformity. The remaining sixty-three patients were observed without any treatment for a mean period of six years and six months (range, one to sixteen years) before skeletal maturity. Of these patients, forty-six eventually had an arthrodesis of the spine, three had not been treated at the time of the most recent follow-up, and fourteen reached skeletal maturity without treatment.

The vertebral anomalies causing the congenital kyphosis or kyphoscoliosis were diagnosed on the basis of anteroposterior and lateral radiographs of the spine. In the first year of life, spinal radiographs of infants could be made only with the infant either supine or sitting, but subsequent radiographs were made with the patient standing. Radiographs of the skeletally immature patients who were observed without treatment were made every four to six months, depending on the severity of the curve. The kyphosis was measured with the modified Cobb method^{4,23} from the most sagittally tilted vertebra at either end of the deformity, as seen on the lateral radiograph. Care was taken to measure all of the serial radiographs from exactly the same anatomical points as were used on the radiograph made when the patient was first evaluated. The senior one of us made the initial measurements on all of the radiographs, and then the other one of us made the measurements again. The radiographs were reviewed together, and the type of vertebral anomaly and the curve measurements were determined through a consensus between us. Intraobserver and interobserver errors were not estimated. Skeletal maturity, reached when there was complete ossification and fusion of the iliac apophysis, was determined radiographically.

Results

The radiographic classification of congenital kyphosis and kyphoscoliosis used in the present series (Table I) is based on the vertebral anomalies that cause the deformity (Fig. 2) and is an expansion of the classification used by Winter et al.²³ We added the subgroups of butterfly (sagittal cleft) and anterior or anterolateral wedged vertebrae, and we differentiated between an anterior and an anterolateral unsegmented bar, as these were not described in the system of Winter et al. The

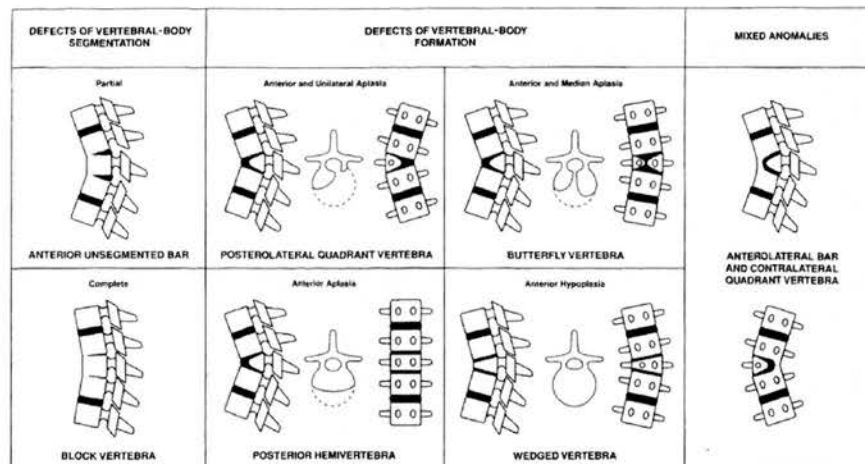


FIG. 2

Drawings showing the different types of vertebral anomalies that produce a congenital kyphosis or kyphoscoliosis.

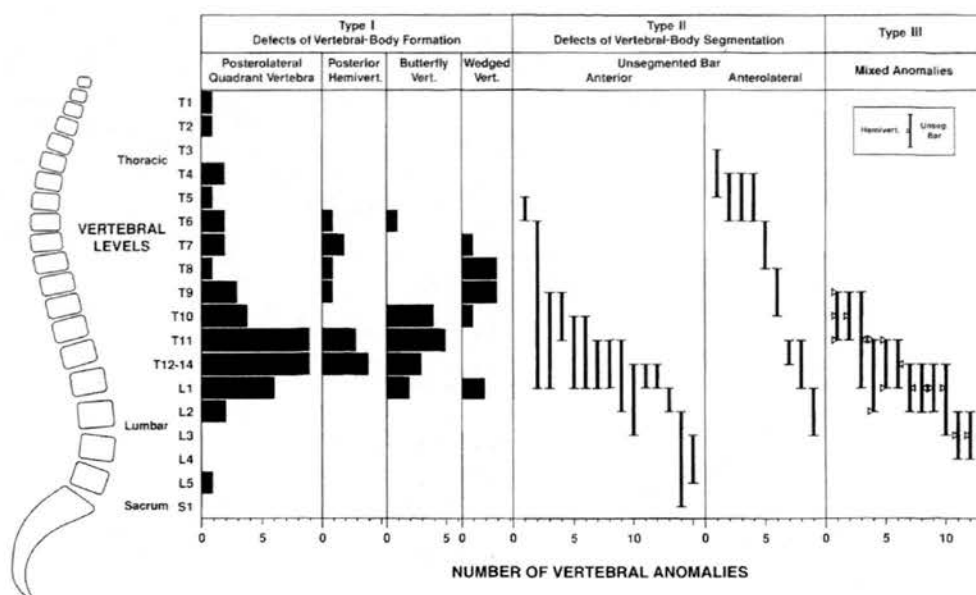


FIG. 3

Graph showing the different types of vertebral defects as well as the number of vertebral levels affected.

number and sites of the vertebral anomalies were assessed (Fig. 3). Thirteen patients had extra thoracic vertebrae. Vertebral anomalies occurred at the thirteenth thoracic level in five patients and at the fourteenth thoracic level in two patients. The present classification system identifies the different types of congenital kyphosis and kyphoscoliosis and can be used to prognosticate about the natural history and the possibility of neurological complications.

The deformity usually progresses more rapidly during the adolescent growth spurt. We estimated the median rate of progression in degrees per year, rather than the mean, for each subgroup of patients: those who were less than ten years old and those who were ten years old and older. We believed that these values were more representative of each subgroup as a whole and were less distorted by widely divergent outliers.

Type I: Anterior Failure of Vertebral-Body Formation

Sixty-eight (61 percent) of the patients had a failure of formation of the anterior segment of the vertebral body. These patients were divided into four subgroups (Table I), depending on the pattern of failure of formation: posterolateral quadrant vertebra, posterior hemivertebra, butterfly (sagittal cleft) vertebra, and anterior or anterolateral wedged vertebra.

Posterolateral Quadrant Vertebra

This anomaly is due to a complete failure of formation of the anterolateral portion of a vertebral body, leaving a posterolateral fragment of bone of varying

size attached to one pedicle and the neural arch (Fig. 2). An anteroposterior radiograph of the spine shows the posterolateral quadrant vertebra as a lateral triangle of bone containing a single pedicle with disc spaces cephalad and caudad (Fig. 4-A). A lateral radiograph shows the varying severity of the anterior failure of formation (Fig. 4-B), which ranges from a near absence of the vertebral body to a tapering of the vertebral body toward the anterior longitudinal ligament.

Thirty-four patients had a single posterolateral quadrant vertebra, and five patients had two adjacent posterolateral quadrant vertebrae. These anomalies occurred at all levels of the spine, but most were at the thoracolumbar junction (Fig. 3), and all patients had a sharply angled kyphoscoliosis. The associated scoliosis was on the left side in twenty-three patients and on the right in sixteen. The mean extent of the measured kyphosis was five vertebrae (range, three to seven vertebrae).

Of the thirty-four patients who had a single posterolateral quadrant vertebra, nineteen were less than ten years old (mean age, three years; range, four months to eight years and seven months) when they were first evaluated; the kyphosis measured a mean of 42 degrees (range, 23 to 88 degrees) at that time. Four patients had an arthrodesis of the spine soon after the initial visit, and the other fifteen patients were observed without treatment for a mean of four years and eight months (range, one year to nine years and two months) before the age of ten years, during which time the median rate of progression of the kyphosis was 2.5 degrees per year (range, 0 to 6 degrees per year) (Figs. 4-A through 4-D). Of the nineteen patients, nine had an arthrodesis before the

age of ten years (mean age, four years and four months; range, nine months to nine years and seven months), when the kyphosis was a mean of 50 degrees (range, 30 to 93 degrees). The other ten patients had not been treated by the age of ten years, at which time the mean kyphosis was 51 degrees (range, 21 to 92 degrees). One of these patients (Case 4, Table II), who had a 92-degree kyphosis at the age of ten years and four months, had a spastic paraparesis. She was managed with a localizer cast and a posterior arthrodesis, and she had a complete recovery.

After the age of ten years, ten patients were seen without treatment for a mean of three years and six months (range, one to six years), during which time the median rate of progression of the kyphosis was 5 degrees per year (range, 2.4 to 12 degrees per year). Ten patients had an arthrodesis of the spine at a mean age of twelve years and eight months (range, eleven years and one month to sixteen years and six months), when the mean kyphosis was 81 degrees (range, 52 to 126 degrees). Two additional patients (Cases 2 and 7, Table II) were seen with a spastic paraparesis at the ages of fourteen years and eight months and sixteen years and ten months, when the kyphosis measured 111 and 60 degrees, respectively. Both patients were managed with an anterior decompression of the spine and arthrodesis combined with a posterior arthrodesis, and they had a complete recovery. Ten patients were seen untreated at skeletal maturity, at which time the mean kyphosis was 62 degrees (range, 40 to 120 degrees). One of the ten patients (Case 6, Table II) was seen with a spastic paraparesis at the age of seventeen years. No treatment was given because of severe congenital cardiac abnormalities, and complete paraplegia developed.

Of the five patients who had two adjacent posterolateral quadrant vertebrae, four were first seen untreated before the age of ten years (mean age, two years and five months; range, nine months to four years and seven months), when the mean kyphosis measured 53 degrees (range, 38 to 73 degrees). One of these patients, who was one year old, had a 50-degree kyphosis and was managed with an immediate arthrodesis of the spine. The other three patients were observed untreated for a mean of six years and ten months (range, five years and five months to nine years and three months) before the age of ten years, during which time the median rate of progression of the kyphosis was 5 degrees per year (4, 5, and 5.5 degrees per year). One of these patients (Case 1, Table II) had a 101-degree kyphosis and a gradual onset of a spastic paraparesis, which was treated with an anterior transthoracic strut-graft arthrodesis and a posterior arthrodesis when she was nine years and two months old. The patient had a partial neurological recovery. The other two patients had an arthrodesis of the spine when they were ten years and twelve years and ten months old and the kyphosis measured 68 and 78 degrees, respectively.

The fifth patient who had two adjacent posterolateral quadrant vertebrae (Case 3, Table II) was seen untreated when he was seventeen years and five months old, at which time he had a 127-degree kyphosis and spastic paraparesis. He was managed with an anterolateral decompression of the spine and a posterior arthrodesis, but he had no improvement and became completely paraplegic.

Posterior Hemivertebra

This anomaly is a complete failure of formation of the anterior portion of the vertebral body, leaving a residual wedged posterior portion attached to both pedicles and the neural arch (Fig. 2). Eight patients had this anomaly: four had a single posterior hemivertebra, and four had two adjacent posterior hemivertebrae. These anomalies all occurred in the caudad thoracic region (Fig. 3) and resulted in a sharply angled kyphosis; any associated scoliosis was less than 20 degrees. The mean extent of the measured kyphosis was five vertebrae (range, four to seven vertebrae).

Two of the four patients who had a single posterior hemivertebra were diagnosed at birth. One patient had a 50-degree kyphosis, which progressed to 54 degrees at one year and was treated with an arthrodesis of the spine. The second patient had a 40-degree kyphosis associated with a forward subluxation of the eleventh thoracic vertebra on the twelfth thoracic vertebra, producing a bayonet-type deformity, and was managed with an arthrodesis of the spine. This deformity, which occurred in only one patient in the present series, has been described as a congenital dislocated spine²⁶ or a congenital vertebral displacement¹⁶. Of the two patients who were not diagnosed at birth, one was three years and six months old and had an 18-degree kyphosis at the initial evaluation. The kyphosis progressed 5 degrees per year to become 55 degrees at the age of ten years and eight months, when the patient had an arthrodesis of the spine. The other patient was seen untreated at skeletal maturity and had a 57-degree kyphosis.

Of the four patients who had two adjacent posterior hemivertebrae, one (Case 5, Table II) was seven years and seven months old at the initial visit and had a 69-degree kyphosis. This kyphosis progressed 9 degrees per year to become 89 degrees at the age of nine years and nine months, at which time the patient had a spastic paraparesis. She had an anterior transthoracic decompression of the spine and arthrodesis combined with a posterior arthrodesis, after which she had increased spasticity. Another patient was eight years and seven months old at the initial visit, at which time she had a 62-degree kyphosis; this progressed 7 degrees per year to become 96 degrees at the age of thirteen years and three months, when she had an arthrodesis of the spine. The remaining two patients were seen untreated at skeletal maturity with kyphoses of 32 and 88 degrees.

TABLE
CLINICAL DATA ON ELEVEN UNTREATED PATIENTS WHO

Case	Type of Congenital Anomaly	Pattern	Site of Vertebral Anomaly	First Evaluation		
				Age (yrs. + mos.)	Curve Magnitude (degrees)	Neurological Status
1	I	Kyphoscoliosis	Posterolateral quadrant (T1 and T2)	3 + 11	73	Normal
2	I	Kyphoscoliosis	Posterolateral quadrant (T4)	14 + 6	104	Normal
3	I	Kyphoscoliosis	Posterolateral quadrant (T5 and T6)	17 + 5	127	Abnormal
4	I	Kyphoscoliosis	Posterolateral quadrant (T7)	2 + 10	50	Normal
5	I	Kyphosis	Posterior hemivertebrae (T6 and T7)	7 + 7	69	Normal
6	I	Kyphoscoliosis	Posterolateral quadrant (T8)	11 + 2	56	Normal
7	I	Kyphoscoliosis	Posterolateral quadrant (T9)	11 + 0	28	Normal
8	IV	Kyphoscoliosis	Unclassifiable	16 + 7	135	Abnormal
9	IV	Kyphoscoliosis	Unclassifiable	1 + 8	99	Normal
10	IV	Kyphosis	Unclassifiable	16 + 6	130	Abnormal
11	IV	Kyphoscoliosis	Unclassifiable	8 + 6	121	Abnormal

*NA = not available.

Butterfly (Sagittal Cleft) Vertebra

This anomaly consists of a partial or complete failure of formation of the anterior and central portions of the vertebral body, leaving two posterolateral fragments of bone attached to the neural arch (Fig. 2). These residual pieces of bone are wedged anteriorly and medially and are separated by a sagittal cleft. This gives the typical butterfly appearance seen on anteroposterior radiographs of the spine, where the wings of the butterfly are represented by the two triangular fragments of the vertebral body, each containing a single pedicle (Fig. 5-A).

All fifteen patients in this subgroup had a single butterfly vertebra. These anomalies occurred mainly at the thoracolumbar junction (Fig. 3) and produced an angular deformity. The posterolateral fragments of bone were symmetrical and produced a pure kyphosis in ten patients, and the fragments were asymmetrical and produced kyphoscoliosis in five patients. The mean extent of the kyphosis was five vertebrae (range, three to seven vertebrae).

Ten patients were first evaluated when they were less than ten years old (mean age, three years and nine

II
HAD SPONTANEOUS NEUROLOGICAL DETERIORATION

Onset of Neurological Deterioration			Treatment	Age at Op. (yrs. + mos.)	Postoperative Neurological Status
Age (yrs. + mos.)	Curve Magnitude* (degrees)	Neurological Status			
9 + 2	101	Gradual onset of spastic paraparesis; normal sensation and bowel and bladder function	Anterior transthoracic strut-graft arthrodesis, posterior arthrodesis	9 + 2	Partial recovery
14 + 8	111	Sudden onset of spastic paraparesis; normal sensation and bowel and bladder function	Anterior transthoracic decompression and strut-graft arthrodesis, posterior arthrodesis	14 + 8	Complete recovery
17 + 5	127	Spastic paraparesis; inability to walk; hypoesthesia; normal bowel and bladder function	Anterolateral decompression, posterior arthrodesis	17 + 5	No recovery, progressed to paraplegia
10 + 4	92	Mild spastic paraparesis; normal sensation and bowel and bladder function	Posterior arthrodesis, localizer cast	10 + 4	Complete recovery
9 + 9	89	Spastic paraparesis; inability to walk; hypoesthesia; urinary difficulty	Anterior transthoracic decompression and arthrodesis, posterior arthrodesis	10 + 0	Increased spasticity
17 + 0	120	Spastic paraparesis; inability to walk; partial bladder and bowel paralysis	No treatment because of severe congenital cardiac abnormalities	—	Progressed to paraplegia
16 + 10	60	Spastic paraparesis; hypoesthesia; normal bowel and bladder function	Anterior transthoracic decompression and arthrodesis, posterior arthrodesis with Luque rods	16 + 10	Complete recovery
16 + 7	135	Spastic paraparesis; paresthesia; normal bowel and bladder function	Anterior transthoracic decompression	16 + 7	Progressed to paraplegia
28 + 1	152	Spastic paraparesis; hypoesthesia; normal bowel and bladder function	Anterior transthoracic strut-graft arthrodesis, posterior arthrodesis	28 + 2	Unchanged, died of respiratory failure
16 + 10	NA	Spastic paraparesis; difficulty walking; hypoesthesia; normal bowel and bladder function	Anterior transthoracic strut-graft arthrodesis, posterior arthrodesis	16 + 10	Partial recovery
8 + 6	121	Sudden onset of spastic paraparesis; inability to walk; hypoesthesia; urinary difficulty	Anterior transthoracic strut-graft arthrodesis, posterior arthrodesis	8 + 6	Partial recovery

months; range, three months to nine years and ten months); the mean kyphosis was 32 degrees (range, 21 to 52 degrees). Seven of the ten patients were observed untreated for a mean of four years and eight months (range, one year and ten months to nine years) before the age of ten years, during which time the median rate of progression of the kyphosis was 1.5 degrees per year (range, 1 to 4 degrees per year). Four patients had an arthrodesis of the spine at a mean age of three years and four months (range, two years and four months to four years and eight months), when the mean kyphosis was 40 degrees (range, 32 to 46 degrees). Six patients had not

been treated by the age of ten years, at which time they had a mean kyphosis of 40 degrees (range, 23 to 58 degrees).

After the age of ten years, five patients were observed untreated for a mean of five years and four months (range, three years to seven years and two months), during which time the median rate of progression of the kyphosis was 4 degrees per year (range, 0 to 12 degrees per year) (Figs. 5-A through 5-D). A sixth patient, who was thirteen years old at the time of the first evaluation, had not been treated and had a 90-degree kyphosis. Six patients had an arthrodesis



FIG. 4-A



FIG. 4-B

Anteroposterior and lateral radiographs of a five-month-old infant, showing a type-I thoracolumbar kyphoscoliosis due to a posterolateral quadrant vertebra at the first lumbar level. The kyphosis measured 38 degrees and the scoliosis, 45 degrees. No treatment was given.

of the spine after the age of ten years (mean age, thirteen years; range, eleven years to fifteen years and six months), when the mean kyphosis was 61 degrees (range, 37 to 90 degrees). Five patients who were seen untreated at skeletal maturity had a mean kyphosis of 47 degrees (range, 23 to 96 degrees).

Anterior and Anterolateral Wedged Vertebrae

These anomalies are characterized by a partial failure of formation, usually affecting the anterolateral segment of the vertebral body (Fig. 2). The anterior wedging always extends forward to the anterior longitudinal ligament, where the anterior height of the wedged vertebra is reduced to between 30 and 50 percent of the posterior height. These anomalies, which were present in six patients, all occurred in the caudad thoracic and cephalad lumbar regions (Fig. 3). The mean extent of the measured kyphosis was six vertebrae (range, three to twelve vertebrae).

A single wedged vertebra was present in two patients: an eleven-year-old who had a 33-degree kyphosis and a seventeen-year-old who had a 43-degree kyphosis. Both of these patients were managed with an arthrodesis of the spine.

There were two adjacent anterolateral wedged vertebrae in four patients. One patient was two years and four months old and had an 85-degree kyphosis, which progressed to 99 degrees by the age of nine years and seven months. The remaining three patients were first seen untreated when they were ten years old or older. One patient had an 85-degree kyphosis at the age of fifteen years and ten months, and two had kyphoses of 90 and 91 degrees at skeletal maturity.

Type II: Anterior Failure of Vertebral-Body Segmentation

Twenty-four patients (21 percent) had an anterior bar of bone extending across the intervertebral disc spaces joining adjacent vertebral bodies. These patients could be divided into two subgroups (Table I) depending on whether the anterior bar of bone lay symmetrically in the sagittal plane, producing a pure kyphosis, or anterolaterally, producing a kyphoscoliosis.

Anterior Unsegmented Bar

This anomaly lay symmetrically in the sagittal plane in fifteen patients; it involved a mean of 3.5 vertebrae (range, two to eight vertebrae) and produced a smooth

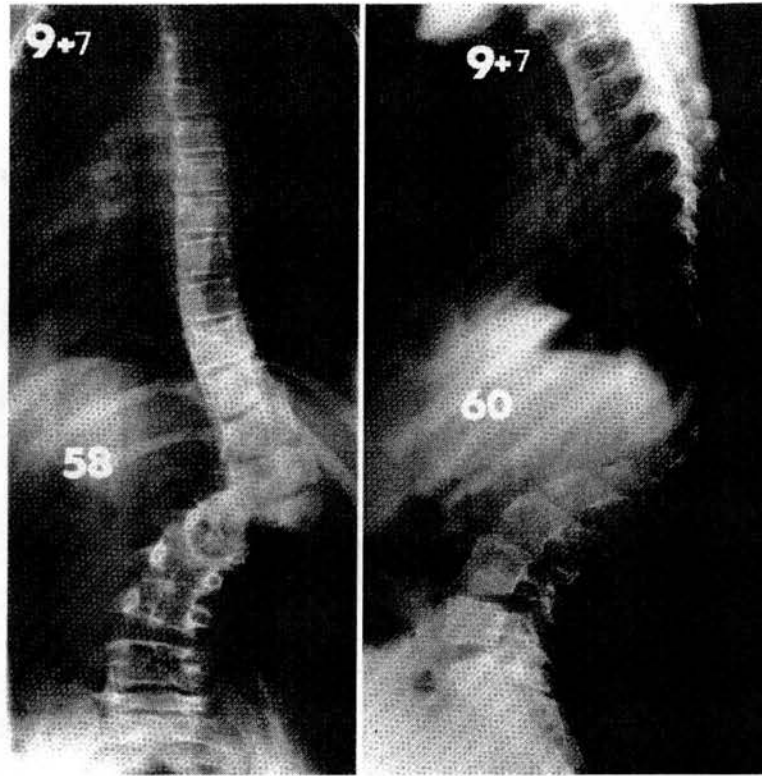


FIG. 4-C

FIG. 4-D

Anteroposterior and lateral radiographs, made when the child was nine years and seven months old, showing a sharp angular deformity. The kyphosis had increased to 60 degrees and the scoliosis, to 58 degrees. The congenital anomaly is seen on both radiographs.

pure kyphosis extending over a mean of five vertebrae (range, three to eleven vertebrae). This anterior failure of segmentation occurred at all levels from the fifth thoracic vertebra to the sacrum (Fig. 3). The anterior-to-posterior depth of the bar relative to the affected disc space was 30 percent in two patients, 50 to 75 percent in twelve patients, and more than 80 percent (a block vertebra) in one patient.

Eight patients were less than ten years old (mean age, five years and two months; range, three months to nine years and five months) when they were first seen, at which time the mean kyphosis was 42 degrees (range, 26 to 48 degrees). Six of these patients were observed without treatment for a mean of eight years and two months (range, five years and three months to nine years and nine months) before the age of ten years, during which time the median rate of progression was 1 degree per year (range, 0 to 1.6 degrees per year). One patient, who was six years old at the time of the initial visit, had an immediate arthrodesis of the spine, when the kyphosis was 67 degrees. The eighth patient, who had a kyphosis of 54 degrees when she was eight years and six months old, was seen only once. Thus, seven of these patients, with a mean kyphosis of 42 de-

grees (range, 23 to 54 degrees), were still untreated by the age of ten years.

Thirteen patients remained untreated after the age of ten years. Six of these patients were less than ten years old when they were first evaluated, and they were observed for a mean of six years and eight months (range, one year and eight months to nine years) after the age of ten years, during which time the median rate of progression of the kyphosis was 1.5 degrees per year (range, 0 to 7 degrees per year) (Figs. 6-A, 6-B, and 6-C). One of the six had an arthrodesis at the age of ten years and one month, one had an arthrodesis at the age of eleven years, and four remained untreated at maturity. Seven of the thirteen patients were first seen when they were at least ten years old. Two of these seven patients had an immediate arthrodesis, one was followed untreated until maturity, and the remaining four were first seen untreated at maturity. The four patients (of the thirteen) who had an arthrodesis were operated on at a mean age of twelve years (range, ten years and one month to fifteen years and four months), when the mean kyphosis was 73 degrees (range, 51 to 90 degrees). The remaining nine patients, who were untreated at skeletal maturity, had

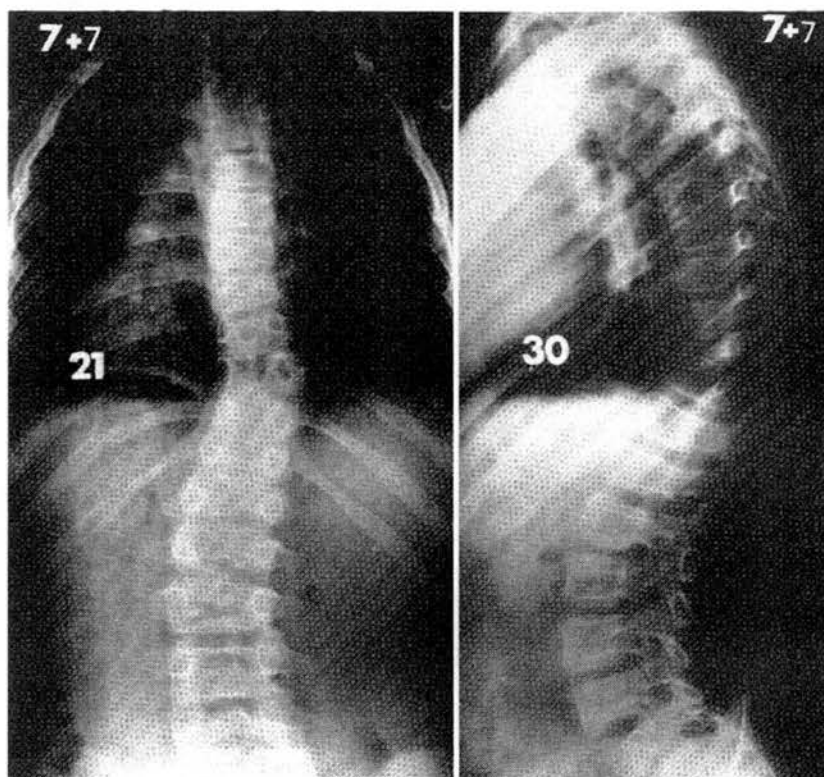


FIG. 5-A

FIG. 5-B

Anteroposterior and lateral radiographs of a child, seven years and seven months old, showing a type-I caudal thoracic kyphoscoliosis due to a butterfly vertebra at the tenth thoracic level. The kyphosis measured 30 degrees and the scoliosis, 21 degrees. No treatment was given.

a mean kyphosis of 64 degrees (range, 27 to 140 degrees) at that time.

Anterolateral Unsegmented Bar

This anomaly, which is asymmetrical and lies anterolaterally, involved a mean of three vertebrae (range, two to three vertebrae) and produced a smooth kyphoscoliosis that extended over a mean of seven vertebrae (range, four to ten vertebrae). The scoliosis was to the right in six patients and to the left in three patients. The anterolateral failure of segmentation affected all levels from the third thoracic vertebra to the third lumbar vertebra (Fig. 3). It was not possible to measure the depth of the bar on the lateral radiographs of the spine because of the kyphoscoliosis.

Two of the nine patients were first seen untreated before they were ten years old. One patient was nine years old and had a 26-degree kyphosis, which was treated with an arthrodesis of the spine. The second patient was six years and ten months old and had a 38-degree kyphosis, which progressed to 42 degrees by the time that the child was ten years old.

Eight patients were seen untreated after the age of ten years. One patient was observed untreated for

four years, during which time the kyphosis progressed 4 degrees per year. Three patients had an arthrodesis of the spine at a mean age of fourteen years (range, thirteen years and ten months to fourteen years and two months), when the mean kyphosis was 90 degrees (range, 65 to 109 degrees). Overall, five of the nine patients were untreated at skeletal maturity, at which time they had a mean kyphosis of 92 degrees (range, 58 to 109 degrees).

Type III: Mixed Anomalies

A combination of an anterolateral unsegmented bar and one or more contralateral posterolateral quadrant vertebrae (Fig. 2) was present in twelve patients (11 percent). The anterolateral bar extended over three adjacent vertebrae in nine patients, four vertebrae in two patients, and five vertebrae in one patient. Nine patients had a single posterolateral quadrant vertebra, two patients had two posterolateral quadrant vertebrae, and one patient had three posterolateral quadrant vertebrae. Localized oblique radiographs of the spine were usually necessary to identify these anomalies, which produced a sharply angular kyphoscoliosis. The anomalies occurred only between the ninth thoracic and the



FIG. 5-C

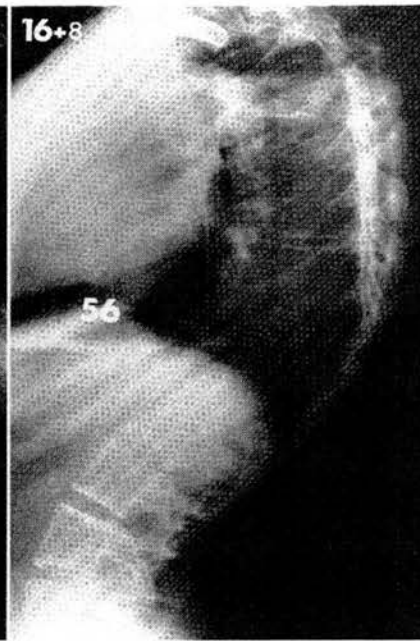


FIG. 5-D

Anteroposterior and lateral radiographs, made when the patient was sixteen years and eight months old, showing that the kyphosis had increased to 56 degrees and the scoliosis, to 46 degrees.

fourth lumbar vertebra (Fig. 3). The degree of the scoliosis, which was to the left in six patients and to the right in six patients, was usually similar to that of the kyphosis. The mean extent of the measured kyphosis was 5.5 vertebrae (range, four to nine vertebrae).

Seven patients were first seen untreated when they were less than ten years old, at a mean age of two years (range, birth to six years), when the mean kyphosis was 29 degrees (range, 11 to 44 degrees). These seven patients were observed untreated for a mean of seven years and two months (range, four years and one month to nine years) before the age of ten years, during which time the median rate of progression of the kyphosis was 5 degrees per year (range, 2.6 to 6 degrees per year). Three patients had an arthrodesis of the spine at a mean age of eight years (range, six years and nine months to nine years and seven months), when the mean kyphosis was 66 degrees (range, 60 to 70 degrees). At the age of ten years, four patients were still untreated and had a mean kyphosis of 76 degrees (range, 70 to 85 degrees).

Three patients were observed untreated for a mean of two years and three months after the age of ten years, during which time the median rate of progression of the kyphosis increased to 8 degrees per year (Figs. 7-A through 7-D). Seven patients had an arthrodesis of the spine at a mean age of thirteen years (range, ten years and seven months to sixteen years and four months), when the mean kyphosis was 92 degrees (range, 85 to

106 degrees). The remaining two patients were seen untreated at skeletal maturity with kyphoses of 60 and 97 degrees.

Type IV: Unclassifiable Anomalies

In eight patients (7 percent), the radiographic characteristics of the congenital anomalies lying at the apex of the curve were obscured by the severity of the deformity; six patients had an angular kyphoscoliosis, and two patients had an angular kyphosis. These curves occurred in all regions of the spine.

Three of the eight patients were first seen between the ages of eight and ten years old. The kyphoses measured 90, 112, and 130 degrees and were treated with an arthrodesis of the spine. A fourth patient (Case 11, Table II), who was eight years and six months old, had a 121-degree kyphosis and a spastic paraparesis. She was managed with an anterior transthoracic strut-graft arthrodesis combined with a posterior arthrodesis and had a partial neurological recovery.

The remaining four patients, who were seen untreated at or near skeletal maturity, had a mean kyphosis of 132 degrees. Three of these patients (Cases 8, 9, and 10, Table II) had a spastic paraparesis when they were between sixteen and twenty-eight years old. Two were managed with an anterior transthoracic strut-graft arthrodesis combined with a posterior arthrodesis; one had a partial recovery, and one had no change in the neurological status and died of respiratory failure. The

third patient had an anterior transthoracic spinal decompression and became paraplegic.

Intraspinal Anomalies

A myelogram or a magnetic resonance imaging scan was made for fifty-one patients either before operative intervention or when there was a spontaneous neurological deficit. Only one of these patients was found to have an occult congenital intraspinal anomaly. This was a ten-year-old boy who had a type-II anterior failure of segmentation extending from the second lumbar vertebra to the sacrum and producing a lumbar kyphosis with its apex at the fourth thoracic vertebra. He had had mild nonprogressive unilateral neurological abnormalities of the lower limb from birth. At the time of exploration, he was found to have a large lumbar extradural lipoma and a tight filum terminale.

Other Congenital Anomalies

Other congenital anomalies were present in thirty-three patients (twenty who had type-I kyphosis or kyphoscoliosis, five who had type-II, and eight who had type-III). None of these anomalies were specific to any one type of congenital kyphosis or kyphoscoliosis.

Anomalies of the lower limb were present in fifteen

patients. Five of these patients had a talipes equinovarus, two had a unilateral cavus foot, and five had mild unilateral hypoplasia of the lower limb. None of these anomalies were associated with a neurological deficit. Other anomalies of the lower limb, each affecting one patient, were unilateral coxa vara and short femur, unilateral slipped capital femoral epiphysis, and syndactyly of the toes.

Cardiac anomalies were diagnosed in five patients; renal anomalies, in four; intestinal abnormalities, in four; esophageal atresia, in two; imperforate anus, in one; and rectovaginal fistula, in one. A Goldenhar syndrome was present in three patients; a scimitar syndrome, in one; and a Prader-Willi syndrome, in one. A Sprengel deformity of the shoulder was found in four patients, and another patient had hypoplasia of the arm.

Discussion

Congenital kyphosis, kyphoscoliosis, and scoliosis form a gradually blending spectrum of spinal deformities that develop due to vertebral anomalies that produce a localized imbalance in the longitudinal growth of the spine. The type of deformity that develops depends on whether the impaired spinal growth occurs unilaterally, producing a pure scoliosis^{9,22}, or is anterior

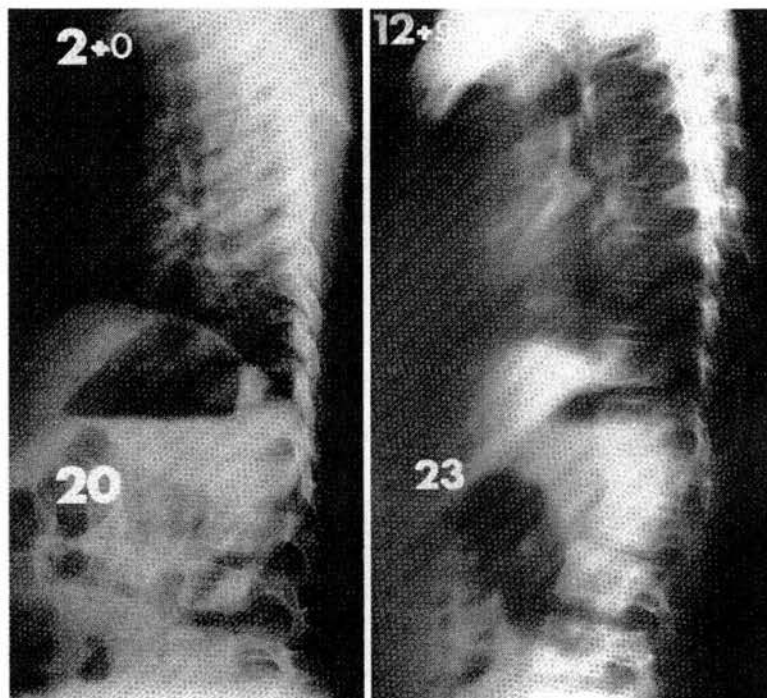


FIG. 6-A

FIG. 6-B

Fig. 6-A: Lateral radiograph of a two-year-old child, showing a type-II thoracolumbar kyphosis due to an anterior unsegmented bar between the twelfth thoracic and the first lumbar vertebra. The kyphosis measured 20 degrees. No treatment was given.

Fig. 6-B: Lateral radiograph, made when the child was twelve years and nine months old, showing that the kyphosis had increased to 23 degrees.

or anterolateral to the transverse axis of vertebral rotation in the sagittal plane, producing a kyphosis or a kyphoscoliosis. In a study of 584 consecutive patients who had congenital spinal deformities, the senior one of us (M. I. McM.) found that 472 patients had a pure congenital scoliosis, seventy-six had a kyphoscoliosis, and thirty-six had a pure kyphosis^{9,12}. In the present study, we looked only at the patients who had a kyphosis or kyphoscoliosis.

We are aware of only one other large study of the natural history of congenital kyphosis and kyphoscoliosis, which was by Winter et al.²³, who reported on 130 patients and classified the kyphoses into three types. Eighty-six (66 percent) of their patients had a type-I kyphosis due to an anterior failure of vertebral-body formation, nineteen patients (15 percent) had a type-II kyphosis due to an anterior failure of vertebral-body segmentation, eighteen patients (14 percent) had a type-III kyphosis in which there was a failure of both formation and segmentation, and seven patients (5 per-

cent) had a kyphosis that could not be classified. However, only thirty of these patients were observed untreated for more than one year (mean, six years; range, one to sixteen years), during which time the mean rate of progression of the kyphosis was 7 degrees per year. The relative rates of progression for the three types of kyphosis could not be determined because too few patients with type-II and type-III anomalies were followed untreated.

In the present study of 112 patients, sixty-three were observed untreated before skeletal maturity for a mean of six years and six months and forty-one were untreated at skeletal maturity. Sixty-eight patients (61 percent) had a type-I kyphosis, twenty-four (21 percent) had a type-II kyphosis, and twelve (11 percent) had a type-III kyphosis (Table I). In eight patients (7 percent), the kyphosis could not be classified because it was seen untreated at a stage when it was so severe that it made it impossible for us to identify the vertebral anomalies precisely (Table I). The ratio of girls to boys was 1.3 to one compared with 1.8 to one in the series of Winter et al.²³. The apex of the kyphosis occurred at all levels from the second thoracic vertebra to the fifth lumbar vertebra (Fig. 8) but was more frequent between the tenth thoracic and the first lumbar vertebra (seventy-four patients; 66 percent). Severe curves occurred at all levels, and we could find no relationship between the severity of the kyphosis and its location in the spine.

We found that progression of the kyphosis or kyphoscoliosis continued throughout growth and usually accelerated during the adolescent growth spurt after the age of ten years before stabilizing at skeletal maturity. The severity and rate of progression of the congenital kyphosis were estimated by measurement, with the modified Cobb method^{4,23}, of serial lateral radiographs of the spine. These measurements represent changes in sagittal inclination that occur at the end vertebrae of the kyphosis, but unfortunately they do not accurately represent the overall curve characteristics or what is happening at the apex of the curve, which is most important²¹. An anterior failure of vertebral-body formation, which produces a sharp angular kyphosis, is much more deforming and potentially neurologically dangerous than a curve, with a similar Cobb measurement, due to an anterior failure of segmentation, which affects several adjacent vertebrae and produces a smooth, less obvious deformity.

In order to understand the natural history of congenital kyphosis and kyphoscoliosis and the disparity in the prognoses for different curves, it is necessary to correlate the principles of normal growth of the spine with the embryological maldevelopment and pathological anatomy of the various types of vertebral anomalies that can cause a kyphosis (Fig. 2). The development of the spine in the embryo occurs rapidly, and the complete anatomical pattern of the vertebrae is formed in mesenchyme during the first six weeks of intrauterine life.

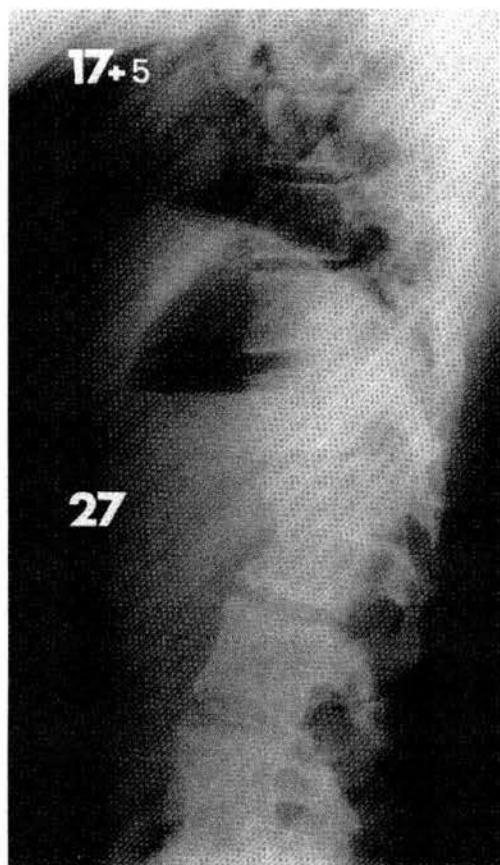


FIG. 6-C

Lateral radiograph, made when the patient was seventeen years and five months old, showing that there had been very little deterioration and that the kyphosis had increased to 27 degrees.

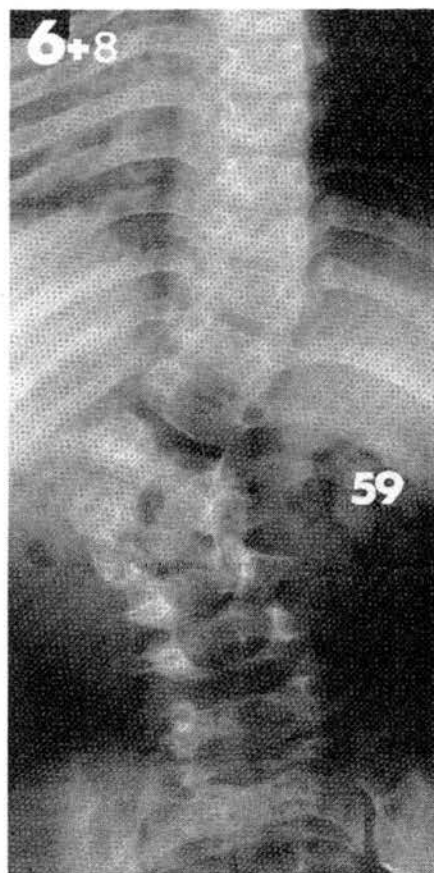


FIG. 7-A



FIG. 7-B

Anteroposterior and lateral radiographs of a child, six years and eight months old, showing a type-III thoracolumbar kyphoscoliosis due to mixed anomalies. There is an anterolateral unsegmented bar extending from the twelfth thoracic to the third lumbar vertebra and a posterolateral quadrant vertebra at the second lumbar level. The kyphosis measured 55 degrees and the scoliosis, 59 degrees. No treatment was given.

Once the mesenchymal mold is established, the cartilaginous and bone stages follow that pattern. The vertebral anomalies that result in the development of a kyphosis are fully established at birth and are thought to develop during the later stages of chondrification and ossification^{17,18}. In contrast, the vertebral anomalies that result in the development of a scoliosis are thought to develop during the earlier mesenchymal stage. Normal longitudinal growth of the spine occurs at the epiphyseal plates on the superior and inferior surfaces of the vertebral bodies¹. A congenital vertebral anomaly that produces a kyphosis has an absence or deficiency of the growth plates lying anterior to the transverse axis of vertebral rotation in the sagittal plane^{17,18}.

The vertebral anomalies causing a type-I kyphosis are thought to be due to a localized failure of vascularization of the developing cartilaginous centrum, resulting in varying degrees of failure of formation of the vertebral body^{15,17,18}. The cartilaginous neural arch develops independent of the centrum and is therefore

not usually affected in this type of anomaly. The most common pattern of failure of vertebral-body formation causing a kyphosis or a kyphoscoliosis in our 112 patients was aplasia of the anterolateral portion of the centrum producing a posterolateral quadrant vertebra (thirty-nine patients; 35 percent). Aplasia of the anterior portion of the centrum alone was much less common and produced a posterior hemivertebra (eight patients; 7 percent). Other anomalies were due to aplasia of the anterior and central portions of the centrum producing a butterfly (or sagittal cleft) vertebra⁶ (fifteen patients; 13 percent) and hypoplasia of the anterior part of the centrum producing an anterior or anterolateral wedged vertebra (six patients; 5 percent). With all of these anomalies, longitudinal growth occurs at the residual epiphyseal plates on the superior and inferior surfaces of the affected vertebral body and in the posterior arch, but there is an absence or deficiency of two epiphyseal plates anteriorly or anterolaterally, resulting in a progressive angular kyphosis or kyphoscoliosis.

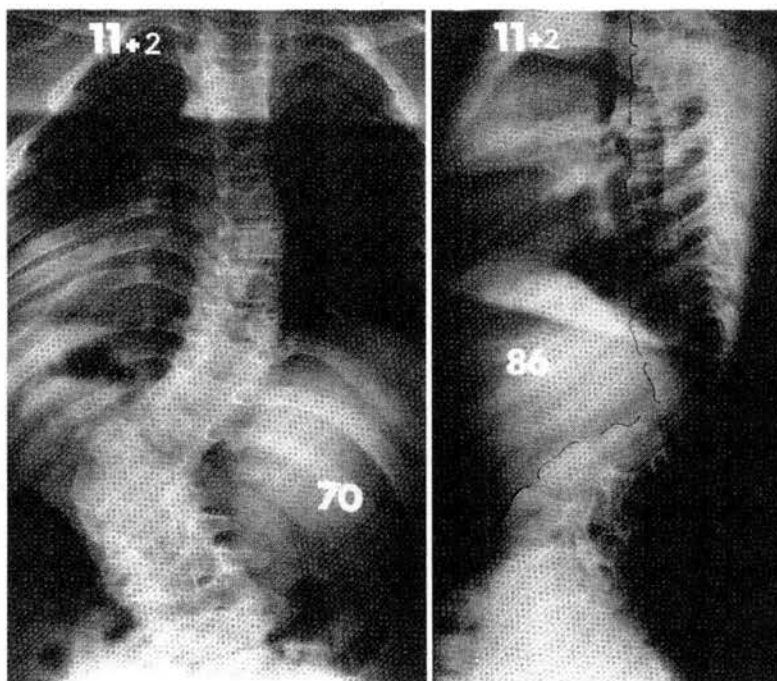


FIG. 7-C

FIG. 7-D

Anteroposterior and lateral radiographs, made when the child was eleven years and two months old, showing that the kyphosis had increased to 86 degrees and the scoliosis, to 70 degrees.

We found that the severity of a type-I kyphosis was proportional to the severity of the anterior failure of formation as shown on the lateral radiograph of the spine. A kyphosis due to a posterolateral quadrant vertebra progressed at a median rate of 2.5 degrees per year before the age of ten years and 5 degrees per year thereafter (Figs. 4-A through 4-D). Ten patients had a spinal procedure at a mean age of twelve years and eight months, when the mean kyphosis was 81 degrees. A kyphosis due to a posterior hemivertebra had only a slightly better prognosis, followed by a butterfly vertebra and a wedged vertebra. A kyphosis due to two adjacent type-I vertebral anomalies progressed more rapidly and produced a more severe deformity than did a similar single anomaly.

Patients with a type-II kyphosis had an unsegmented bar of bone extending anteriorly across the intervertebral disc spaces, joining a mean of 3.5 vertebrae (Fig. 2). This type of anomaly is thought to be due to bone metaplasia occurring in the anterior part of the annulus fibrosus and ring apophysis during the late chondrification and ossification periods^{14,17,18}. Longitudinal growth is impaired anteriorly where the osseous bar has replaced the epiphyseal plates on the superior and inferior surfaces of the vertebral bodies, whereas some degree of growth occurs posteriorly cephalad and caudad to the residual discs and in the neural arch. However, we could find no constant relationship be-

tween the size of the kyphosis and the number of vertebrae involved or the degree of disc-space involvement. These patients had a variable prognosis depending on whether the unsegmented bar of bone lay symmetrically in the sagittal plane or anterolaterally. In fifteen patients (13 percent), the bar of bone lay symmetrically, producing a pure kyphosis; although this deformity could be large, it usually progressed relatively slowly at a median rate of 1 degree per year before the age of ten years. Only two patients who were followed after the age of ten years had a kyphosis that progressed at a rate of more than 2 degrees per year. Nine patients were untreated at skeletal maturity, and at that time the mean kyphosis was 64 degrees. An anterolateral bar producing a kyphoscoliosis occurred in only nine patients (8 percent) but had a much worse prognosis. In five patients who were seen untreated at skeletal maturity, the mean kyphosis was 92 degrees. Morin et al.¹⁴ followed ten patients who had a type-II kyphosis and found, at a mean of six years, that only one curve had progressed more than 1 degree per year. In contrast, Mayfield et al.¹³ reported that eight patients with a type-II kyphosis followed for a mean of six years had a mean rate of progression of 5 degrees per year.

A type-III kyphoscoliosis due to an anterolateral unsegmented bar combined with contralateral posterolateral quadrant vertebrae was least common, occurring in only twelve (11 percent) of the 112 patients, but it

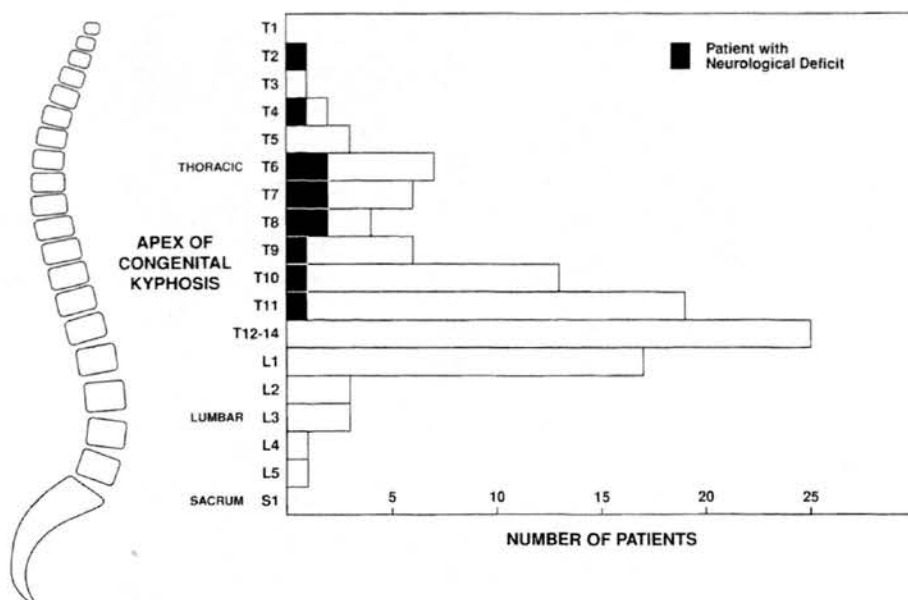


FIG. 8

Histogram showing the level of the apex of the congenital kyphosis or kyphoscoliosis in the 112 patients and the number of patients who had a spontaneous neurological deficit.

usually progressed the most rapidly and produced the most severe deformity (Figs. 7-A through 7-D). This type of kyphosis progressed at a median rate of 5 degrees per year before the age of ten years and 8 degrees per year thereafter. Seven patients had an arthrodesis of the spine at a mean age of thirteen years, when the mean kyphosis was 92 degrees.

A progressive spastic paraparesis of the lower limbs due to anterior compression of the spinal cord at the apex of the congenital kyphosis or kyphoscoliosis occurred spontaneously in eleven (10 percent) of the 112 patients, all of whom had been neurologically normal previously (Table II). Seven of these patients had a type-I anomaly (four had a single posterolateral quadrant vertebra, two had two adjacent posterolateral quadrant vertebrae, and one had two adjacent posterior hemivertebrae), and four patients had anomalies that could not be classified because of the severity of the angular kyphosis. Neurological complications did not occur in patients with recognizable type-II anomalies because they produced a smooth kyphosis in which the abnormal vertebrae were stabilized by the anterior failure of segmentation. However, a posterolateral quadrant vertebra and a posterior hemivertebra were unstable and tended to extrude backward into the spinal canal, causing anterior compression of the spinal cord at the apex of the angular deformity as it became more severe. It is likely that many more patients in the series would have had spontaneous neurological deterioration if they had not had a spinal arthrodesis at an earlier stage. The apex of the kyphosis in nine of the eleven patients who had

neurological complications was in the middle and caudal thoracic regions (Fig. 8), where the diameter of the spinal canal is narrowest and the spinal cord has a relatively poor blood supply. A congenital kyphosis or kyphoscoliosis with its apex cephalad to the fifth thoracic vertebra was uncommon, but two of four patients with a curve in this region had a neurological deficit. No patient in whom the apex of the kyphosis was at or caudal to the twelfth thoracic vertebra had neurological abnormalities. The onset of neurological deterioration occurred between the ages of eight and eleven years in four patients, between the ages of fourteen and eighteen years in six, and at twenty-eight years in one. The mean size of the kyphosis at the onset of paraparesis was 111 degrees, but one patient who had a posterolateral quadrant vertebra at the ninth thoracic level had only a 60-degree kyphosis. Winter et al.²³ found that sixteen (12 percent) of the 130 patients in their series became paraparetic at a mean age of twelve years (range, four to nineteen years) and paraparesis occurred only with type-I anomalies, usually in the cephalad thoracic region.

Fifty-one of our patients had a myelogram or a magnetic resonance imaging scan. However, only one of them was found to have an occult congenital intraspinal anomaly. This patient had a lumbar extradural lipoma and a tight filum terminale in association with a lumbosacral anterior failure of segmentation. It is possible that more occult intraspinal anomalies would have been identified if all 112 of our patients had had a magnetic resonance imaging scan. To our knowledge, the only other report of intraspinal anomalies occurring in asso-

ciation with a congenital kyphosis was by Bradford and Kahmann³, who described four patients who had a posterior hemivertebra at the fifth lumbar level producing a lumbosacral kyphosis in association with diplomyelia and a tight filum terminale. The rarity of intraspinal anomalies in our patients contrasts with the reported prevalence of intraspinal anomalies in congenital scoliosis, which has ranged from 6 percent (twenty-three of 392 patients)²⁴ to 58 percent (sixty-three of 108 patients)², with a diastematomyelia being most common. A possible explanation for this disparity is that the vertebral anomalies responsible for a congenital scoliosis appear during the mesenchymal period, when the spinal cord is also developing, whereas the anomalies responsible for a congenital kyphosis or kyphoscoliosis occur during the late chondrification and ossification periods, when the basic anatomy of the neural structures has already been established^{17,18}.

In conclusion, congenital kyphosis and kyphoscoliosis are uncommon but potentially dangerous spinal deformities that, in contrast to congenital scoliosis, can

occasionally result in paraplegia. The key to successful management is to recognize the poor prognosis at an early stage, preferably before the child is five years old, and to balance spinal growth by means of a simple posterior arthrodesis of the spine before the kyphosis exceeds 45 degrees²⁵. This allows progressive reduction of a type I kyphosis or kyphoscoliosis due to continuing anterior longitudinal growth in the presence of the posterior tether produced by the arthrodesis. Type-II and type III deformities, which involve an anterior failure of vertebral body segmentation, have no potential for anterior growth and therefore are only stabilized by a posterior arthrodesis. Delaying operative intervention until a later stage necessitates much more complex and potentially hazardous anterior and posterior procedures on the spine to correct and stabilize the deformity. A thorough knowledge of the natural history is essential in planning treatment and preventing neurological complications.

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The Surgical Management of Congenital Kyphosis and Kyphoscoliosis

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Study Design. A retrospective study of surgery for congenital kyphosis and kyphoscoliosis.

Objective. To assess the effectiveness of different types of spine surgery in the management of congenital kyphosis and kyphoscoliosis.

Summary of Background Data. Congenital kyphosis and kyphoscoliosis are much less common than congenital scoliosis but potentially more serious, because these curves can progress rapidly and Type I deformities can lead to spinal cord compression and paraplegia. No one operative procedure can be applied to all types and sizes of deformity. The method of surgical treatment depends on the age of the patient, the type and size of the deformity, and the presence or absence of spinal cord compression causing a neurologic deficit.

Methods. Sixty-five patients with a congenital kyphosis ($n = 14$) or kyphoscoliosis ($n = 51$) were treated by five different methods of spine arthrodesis: prophylactic posterior arthrodesis before age of 5 years ($n = 11$), posterior arthrodesis after age 5 years without instrumentation ($n = 26$) and with instrumentation ($n = 12$), combined anterior and posterior arthrodesis without instrumentation ($n = 7$) and with instrumentation ($n = 9$). Six patients had preoperative lower limb spastic paraparesis caused by spinal cord compression. The mean age at surgery was 9 years 6 months (range, 11 months to 25 years), and all 65 patients were observed for a minimum of 2 years (mean 6 years 6 months, range 2 to 18 years). Fifty-seven patients reached skeletal maturity.

Results. A posterior arthrodesis performed before the age of 5 years resulted in a gradual reduction of the kyphosis by a mean 15° in 9 of the 11 patients, followed up for a mean of 11 years, whose initial kyphosis was less than 55° . Patients treated after the age of 5 years by a posterior arthrodesis followed by cast application had poor correction and a high incidence of pseudarthrosis. This was not significantly improved by the addition of posterior instrumentation. For curves greater than 60° , the most successful results were achieved by an anterior spinal release and arthrodesis with strut graft correction followed by posterior arthrodesis with instrumentation (if possible).

Conclusion. All patients with a Type I or Type III congenital kyphosis or kyphoscoliosis should be treated by a posterior arthrodesis before the age of 5 years and before the kyphosis exceeds 50° . A kyphosis that does not reduce to less than 50° as measured on the lateral spine radiograph made with the patient supine requires an anterior release and arthrodesis with strut grafting followed

by posterior arthrodesis with instrumentation (if possible). [Key words: surgery, congenital kyphosis] *Spine* 2001;26:2146–2155

Congenital kyphosis or kyphoscoliosis is an uncommon deformity in which there is an abnormal posterior convex angulation of a segment of the spine caused by developmental vertebral anomalies that impair longitudinal growth anterior or anterolateral to the transverse axis of vertebral rotation in the sagittal plane.¹⁰ These curves have been classified into those caused by anterior failure of vertebral body formation (Type I), anterior failure of vertebral body segmentation (Type II), and a mixture of failure of formation and segmentation (Type III).^{10,16} These types of spine deformity are much less common than congenital scoliosis^{7–9,15} but are potentially more serious because a Type I deformity can lead to spinal cord compression and paraplegia.^{4,10,16}

Brace treatment is ineffective,^{4,16} and surgical treatment is frequently necessary, but few large series have been reported.^{6,11,16–18} However, no one operative procedure is appropriate for all types and sizes of deformity. The method of surgery depends on the age of the patient, the type of vertebral anomaly, the size of the deformity, and the presence or absence of spinal cord compression. Successful surgery depends on selecting the right procedure and applying it at the appropriate time.

The purpose of this article is to assess the effectiveness of different types of spine surgery in the management of congenital kyphosis and kyphoscoliosis. Prophylactic surgery was applied to young patients presenting before the age of 5 years with small curves that had a poor prognosis. The objective of surgery was to prevent further progression and, if possible, achieve gradual correction by balancing the growth of the spine. Patients presenting after the age of 5 years were treated by either posterior or combined anterior and posterior surgery with or without spine instrumentation.

Materials and Methods

The authors retrospectively reviewed the medical records and spine radiographs of 69 consecutive patients with a congenital kyphosis or kyphoscoliosis treated by a spine arthrodesis at the Princess Margaret Rose Orthopaedic Hospital (Edinburgh, UK) between 1965 and 1996. Patients who had a kyphosis associated with myelomeningocele, Scheuermann's disease, neurofibromatosis, skeletal dysplasia, infection, or trauma were excluded. Four patients who were followed up for less than 2 years were also excluded. The remaining 65 patients were observed for a mean 6 years 6 months (range, 2 years to 18 years),

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Conflict of interest category: 12.

Table 1. Clinical Data on 65 Patients With Congenital Kyphosis and Kyphoscoliosis Treated Surgically

Type of Surgical Procedure	No. of Patients	At Surgery			At Final Follow-up		
		Age: Mean: y + mo (Range)	Size of Kyphosis: Mean Degree (Range)	Extent of Fusion: Mean no. of Vertebrae	Postop Correction: Mean Degrees (Range)	Loss in Correction at 1 yr: Mean Degrees (Range)	Size of Kyphosis: Mean Degree (Range)
Posterior arthrodesis without instrumentation before age 5 y	11	2 + 3 (0 + 11 to 4 + 8) SD 1 + 2	43° (30 to 55) SD 9°	5 (4 to 7) SD 1	4° (0 to 10) SD 3°	4° (0 to 19) SD 6°	35° (5 to 81) SD 21°
Posterior arthrodesis without instrumentation after age 5 y	26	12 + 0 (6 + 8 to 25) SD 3 + 6	70° (26 to 128) SD 25°	7 (4 to 11) SD 2°	9° (0 to 33) SD 9°	6° (0 to 22) SD 5°	72° (29 to 132) SD 26°
Posterior arthrodesis with instrumentation	12	12 + 2 (6 + 9 to 17 + 4) SD 3 + 4	62° (33 to 88) SD 20°	8 (3 to 10) SD 2	17° (3 to 40) SD 12°	10° (0 to 36) SD 8°	61° (29 to 81) SD 19°
Combined anterior and posterior arthrodesis without instrumentation	7	12 + 6 (9 + 3 to 16 + 6) SD 2 + 11	97° (83 to 119) SD 13°	8 (7 to 12) SD 2	18° (3 to 30) SD 10°	2° (0 to 5) SD 2°	78° (60 to 98) SD 15°
Combined anterior and posterior arthrodesis with instrumentation	9	13 + 8 (9 + 7 to 19) SD 3 + 2	90° (62 to 126) SD 20°	11 (8 to 14) SD 2°	32° (14 to 53) SD 14°	3° (0 to 15) SD 4°	59° (45 to 82) SD 13°
							Duration: Mean: y + mo (Range)
							11 + 0 (2 + 6 to 18) SD 4 + 4
							5 + 3 (2 + 0 to 9 + 10) SD 2 + 5
							4 + 6 (2 + 3 to 10 + 0) SD 2 + 7
							5 + 4 (3 + 0 to 8 + 2) SD 1 + 11
							4 + 3 (2 + 0 to 8 + 5) SD 2 + 5
							Comment
							Kyphosis improved >10° 9 patients Loss in correction >10° 2 patients Pseudarthrosis 6 patients Loss in correction >10° 13 patients Pseudarthrosis 2 patients Loss in correction >10° 7 patients All skeletally mature with solid stable fusion All skeletally mature with solid stable fusion

and 57 patients reached skeletal maturity. Fifty-five of the most recent patients were operated on by the senior author (M.J.M.).

There were 36 female and 29 male patients, whose mean age at operation was 9 years 6 months (range, 11 months to 25 years). The deformity was classified as Type I (anterior failure of vertebral body formation) in 46 patients, Type II (anterior failure of vertebral body segmentation) in eight patients, or Type III (mixture of failure of formation and segmentation) in 10 patients. One patient could not be classified because the vertebral anomalies were partially obscured by the severity of the deformity. No patient had a congenital dislocated spine or a congenital vertebral displacement, which are extremely rare deformities usually diagnosed shortly after birth.^{5,13,19}

Kyphoscoliosis was present in 51 patients and pure kyphosis in 14 patients. In patients with kyphoscoliosis, the kyphosis was always the major component of the deformity and exceeded 25° measured over the anomalous segment. The apex of the deformity occurred at all levels of the spine from T2 to L4 but was most frequent between T10 and L1 inclusive (45 patients, 69%). The mean extent of the kyphosis was over five vertebrae (range, 3–9).

Before surgery, 35 patients had a myelogram and four had MRI of the spine. Six of these patients presented before surgery with spastic paraparesis, and all were found to have anterior spinal cord compression at the apex of the kyphosis or kyphoscoliosis. No patient had an occult congenital intraspinal anomaly. The authors' present policy is to perform MRI on all patients with congenital kyphosis or kyphoscoliosis before spine surgery.

The severity of the kyphosis was measured in all patients by use of the modified Cobb method^{3,16} from the most sagittally tilted vertebra at either end of the deformity as seen on the lateral spine radiographs taken with the patient erect immediately before surgery, after surgery, at 1 year, and at final follow-up. The scoliosis, when present, was measured by use of the Cobb method³ on the anteroposterior spine radiographs. Care was taken to measure all the radiographs from exactly the same anatomic points with reference to the preoperative radiograph. The senior author made the initial measurements on all of the radiographs, and the second author made the measurements again. The radiographs were reviewed together, and the curve measurements determined through a consensus between the authors. Intraobserver and interobserver errors were not established. Unfortunately, anteroposterior and lateral spine radiographs often fail to fully represent the full severity of the three-dimensional deformity in patients with severe kyphoscoliosis. Skeletal maturity was determined radiographically when there was complete ossification and fusion of the iliac apophyses.

The surgery was judged to have failed if at final follow-up the kyphosis was 10° or more than it was immediately after the surgery or if there was no neurologic improvement in patients with spinal cord compression. A Mann-Whitney *U* test was used to test the magnitude of mean change in kyphosis angle between groups, a *P* value of 0.05 being considered significant.

Results

There were five different methods of spine arthrodesis (Table 1) depending on the age of the patient, the type of vertebral anomaly, and the size of the deformity. In addition, of the six patients with spinal cord compression and spastic paraparesis (Table 2), three had a direct an-

Table 2. Treatment of Six Patients With Preoperative Neurologic Deficit

Type of Congenital Anomaly	Pattern	Preoperative		Neurologic Status	Final Follow-up					
		Site of Vertebral Anomaly	Age at Operation (y + mo)		Kyphosis (°)	Correction Obtained (°)	Age (y + mo)	Kyphosis (°)	Neurologic Status	
1	Kyphoscoliosis	Posterolateral quadrant (T7)	10 + 4	92	Mild spastic paraparesis, able to walk, normal sensation and bowel and bladder function	(1) Posterior arthrodesis T3–T10 without instrumentation (2) Explored at 6 months; solid posterior fusion	8	17 + 8	98	Complete recovery over 1 mo
1	Kyphoscoliosis	Posterolateral quadrant (T5 and T6)	17 + 5	127	Spastic paraparesis, inability to walk, hypoesthesia, normal bladder and bowel function	(1) Costotransversectomy and anterior spine decompression (2) Posterior arthrodesis T2–T9 without instrumentation	Not known	32 + 4	127	Initial improvement to mild spasticity, but at age 32 y gradual deterioration to paraplegia
1	Kyphoscoliosis	Posterolateral quadrant (T9)	16 + 10	60	Spastic paraparesis, difficulty walking, hypoesthesia, normal bowel and bladder function	(1) Costotransversectomy and anterior spine decompression (2) Posterior arthrodesis T5–L1 with Luque instrumentation	20	19 + 1	45	Complete recovery over 3 mo
1	Kyphoscoliosis	Posterolateral quadrant (T4)	14 + 8	111	Sudden onset of spastic paraparesis, inability to walk, normal sensation and bowel and bladder function	(1) Anterior transthoracic strut graft arthrodesis T1–T8 (2) Posterior arthrodesis T1–T9 without instrumentation	30	18 + 11	84	Complete recovery over 6 mo
1	Kyphoscoliosis	Posterolateral quadrant (T1 and T2)	9 + 2	101	Gradual onset of spastic paraparesis, difficulty walking, normal sensation and bowel and bladder function	(1) Anterior transthoracic strut graft arthrodesis C7–T5 (2) Posterior arthrodesis C6–T5 without instrumentation	21	18 + 2	82	Partial recovery over 6 months, residual spasticity but able to walk, normal sensation and bowel and bladder function
1	Kyphosis	Posterior hemivertebrae (T6 and T7)	9 + 9	89	Spastic paraparesis, inability to walk, hypoesthesia, urinary difficulty	(1) Anterior transthoracic decompression and arthrodesis (2) Posterior arthrodesis T4–T10 without instrumentation (3) Explored at 6 months; solid posterior fusion	6	18 + 2	96	Complete motor paralysis, but sensation and bowel and bladder function normal

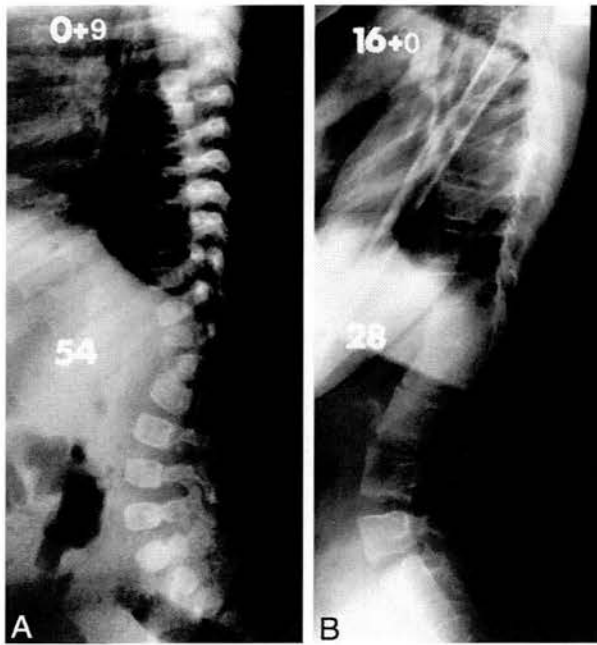


Figure 1. **A**, Infant aged 9 months with a congenital thoracolumbar kyphosis caused by a posterior hemivertebra at T12. A posterior arthrodesis was performed when the infant was 11 months old and the kyphosis measured 54°. **B**, When the patient was aged 16 years, the congenital kyphosis had improved to 28° and there was no cosmetic deformity.

terior decompression and the remaining three had an indirect decompression by reducing the deformity.

Prophylactic Posterior Spine Arthrodesis Before the Age of 5 Years

There were 11 patients in this group, and all had a Type I deformity caused by anterior failure of vertebral body formation. No patient had a neurologic deficit. The object of surgery was to prevent further progression and, if possible, achieve gradual correction by balancing the growth of the spine.

All 11 patients underwent posterior arthrodesis by excision of the facet joints and decortication of the posterior vertebral structures followed by an onlay of allograft bank bone from degenerate femoral heads. These patients were too young to have autograft bone taken from the iliac crest. After surgery, an underarm plaster jacket was worn for 6 months.

The mean age at surgery was 2 years 5 months (range, 11 months to 4 years 8 months), and the mean kyphosis was 43° (range, 30–55°). The arthrodesis extended over a mean five vertebrae (range, 4–7). After surgery, the patients were observed for a mean 11 years (range, 3 years 6 months to 18 years), and during this period the kyphosis slowly improved in nine of the 11 patients by a mean of 15° (range, 11–25°) (Figure 1). Five patients reached skeletal maturity.

Two patients had further progression of their deformity. One of these patients underwent posterior arthrodesis at the age of 3 years 4 months when the kyphosis was 36°. Postoperatively, the kyphosis slowly pro-

gressed, and it measured 50° when the patient was 5 years 6 months old. At this time a pseudarthrosis was repaired posteriorly and reinforced by an anterior rib strut graft. She was followed up for an additional 4 years with no further progression. The second patient underwent posterior arthrodesis at the age of 4 years 8 months when the kyphosis was 45°. Initially, the curve remained stable, as measured on 6-monthly serial spine radiographs, until the onset of the adolescent growth spurt at 12 years, when further normal vertebrae were added above and below the fused segment and the overall kyphosis measured 84°. At this time the posterior fusion, which was found to be solid, was extended to two levels above and two levels below the previous fusion and reinforced by an anterior vascularized rib strut graft. She was followed up to skeletal maturity, when the kyphosis measured 84°.

Posterior Spine Arthrodesis with Cast Correction After the Age of 5 Years

There were 26 patients in this group. The deformity was Type I in 17 patients, Type II in 6 patients, Type III in 2 patients, and unclassified in 1 patient. Before surgery, 2 patients (Table 2 Patients 1 and 2) with Type I deformities had spinal cord compression and lower limb spastic paraparesis.

All 26 patients underwent posterior arthrodesis by excision of the facet joints followed by decortication and an onlay of autogenous iliac crest bone grafts. One of the patients (Patient 2) who had a neural deficit also underwent costotransversectomy and anterior decompression of the spinal cord. After surgery, all the patients had a modified Risser-type localizer jacket applied while awake on a traction table. The jacket was worn for 6 to 9 months.

The mean age of the 26 patients at posterior spine arthrodesis was 12 years (range, 6 years 8 months to 25 years) when the mean kyphosis was 70° (range, 26–128°). The mean extent of the kyphosis was over five vertebrae (range, 3–8), and the posterior arthrodesis was over seven vertebrae (range, 4–11). After surgery, the kyphosis was corrected by only a mean 9° (range, 0–33°) in a spinal jacket. One patient who before surgery was neurologically normal was noted after surgery to have a spastic paraparesis, which slowly improved over 6 months but did not fully recover. At 6 months, 13 patients underwent planned exploration of their fusion, and six were found to have a pseudarthrosis, which was repaired. The mean loss in correction at 1 year was 5° (range, 0–19°).

The 26 patients were observed for a mean 5 years 3 months (range, 2 years to 9 years 10 months), and 25 patients reached skeletal maturity. At this time the mean kyphosis was 72° (range, 29–132°). Thirteen patients had lost 10° or more correction (mean 19°, range 10–50°), and nine of them had a kyphosis greater than it was before surgery. The remaining 13 patients had lost less

than 10° of correction, and four of them had spontaneously improved by 3 to 6°.

Of the two patients who had a preoperative neurologic deficit, one patient (Patient 1) who had mild spasticity made a complete recovery over 1 month while in a spinal jacket, and the condition of the other patient (Patient 2), who had a more severe neurologic deficit and had been treated by a costotransversectomy and anterior spinal cord decompression, improved to mild spasticity but later deteriorated to complete paraplegia when the patient was 32 years of age (Table 1).

Posterior Spine Arthrodesis With Instrumentation

There were 12 patients in this group. The deformity was Type I in nine patients and Type III in three patients. Before surgery, one patient (Table 2, Patient 3) with a Type I deformity had spinal cord compression and a lower limb spastic paraparesis.

All the patients underwent posterior arthrodesis with autogenous iliac crest bone grafts and correction of the curve by posterior instrumentation. These patients were treated at various times over the period of review, during which time the method of instrumentation evolved: Harrington instrumentation in 10 patients, Luque instrumentation with sublaminar wires in one patient, and AO universal spine system instrumentation in one patient. The one patient (Patient 3) with a preoperative neural deficit also underwent costotransversectomy and anterior decompression of the spinal cord. After surgery, an underarm plaster jacket was applied to all patients and worn for 6 months.

The mean age of these 12 patients at surgery was 12 years 2 months (range, 6 years 9 months to 17 years 4 months), when the mean kyphosis was 62° (range, 33–88°). The mean extent of the kyphosis was over five vertebrae (range, 3–9), and the arthrodesis was over eight vertebrae (range, 3–10). After surgery, the kyphosis was corrected by a mean 17° (range, 3–40°).

Two patients, one aged 9 years 7 months and the other aged 11 years 8 months, with kyphosis of 60° and 33°, respectively, were treated by Harrington instrumentation, which broke. Their curves progressed to 87° and 63° when they were 11 years 3 months old and 15 years 2 months old, respectively. At this time, both patients underwent posterior repair of a pseudarthrosis, and the patient with the larger curve had an additional reinforcement by an anterior arthrodesis and rib strut graft. After this there was no further loss in correction, and both patients were observed to skeletal maturity.

The mean loss in correction in the remaining 10 patients at 1 year was 7° (range, 0–14°). These patients were observed for a mean 4 years (range, 2 years to 10 years), and nine patients reached skeletal maturity. At this time the mean kyphosis was 58° (range, 32–81°). Five patients had lost 10° or more of correction (mean 18°, range 14–24°), and three of these patients had a kyphosis greater than it was before surgery.

The one patient (Patient 3) with a preoperative neural deficit made a complete recovery over 3 months.

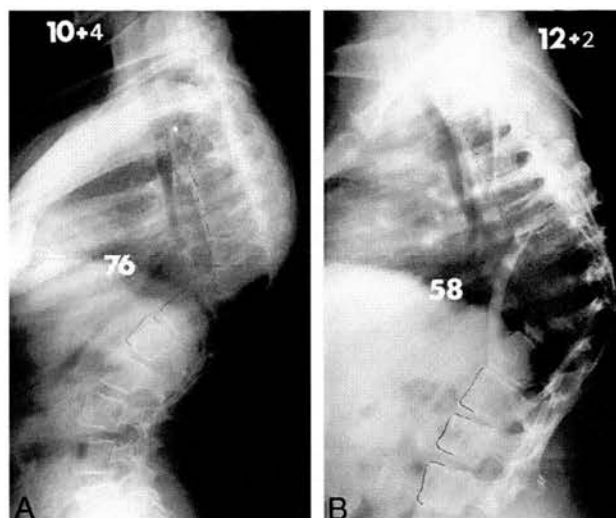


Figure 2. **A**, Girl aged 10 years 4 months with a 76° congenital thoracic kyphosis caused by a posterior hemivertebra at T11. She was treated by an anterior release and vascularized rib strut graft combined with a posterior arthrodesis. **B**, When the patient was 12 years 2 months old, there was a solid fusion with hypertrophy of the anterior rib strut graft. The congenital kyphosis was stable at 58°.

Combined Anterior and Posterior Spine Arthrodesis Without Instrumentation

There were seven patients in this group. The deformity was Type I in five patients and Type II in two patients. Before surgery, three patients (Table 2, Patients 4, 5, and 6) with Type I deformities had spinal cord compression and a lower limb spastic paraparesis.

All seven patients underwent two-stage anterior and posterior spine surgery. The five patients who had anterior failure of vertebral body formation (Type I deformity) underwent first-stage thoracotomy and anterior spinal release by multiple disc excisions followed by partial correction of the deformity by use of rib (3 patients) or fibular (1 patient) strut grafts or a vascularized rib strut graft (1 patient) (Figure 2). One of the patients with a preoperative neurologic deficit (Patient 6) also underwent anterior transthoracic decompression of the spinal cord at the apex of the deformity. In the two patients with anterior failure of vertebral segmentation (Type II deformity), a first-stage anterior approach to the spine was used to perform osteotomies of the unsegmented bar and disc excisions followed by partial correction by use of rib strut grafts. After 5 days, all seven patients underwent second-stage posterior arthrodesis by use of autogenous iliac crest bone grafts. It was not possible to apply posterior spine instrumentation to any of these seven patients, either because of the small size of their vertebrae or because of the severity of their deformity. After surgery, a modified localizer jacket was applied while the patient was under traction and awake, and worn for 6 to 9 months.

The mean age of the patients at surgery was 12 years 6 months (range, 9 years 3 months to 16 years 6 months), and the mean kyphosis was 97° (range, 83–119°). The mean extent of the kyphosis was over six vertebrae (range, 4–8),

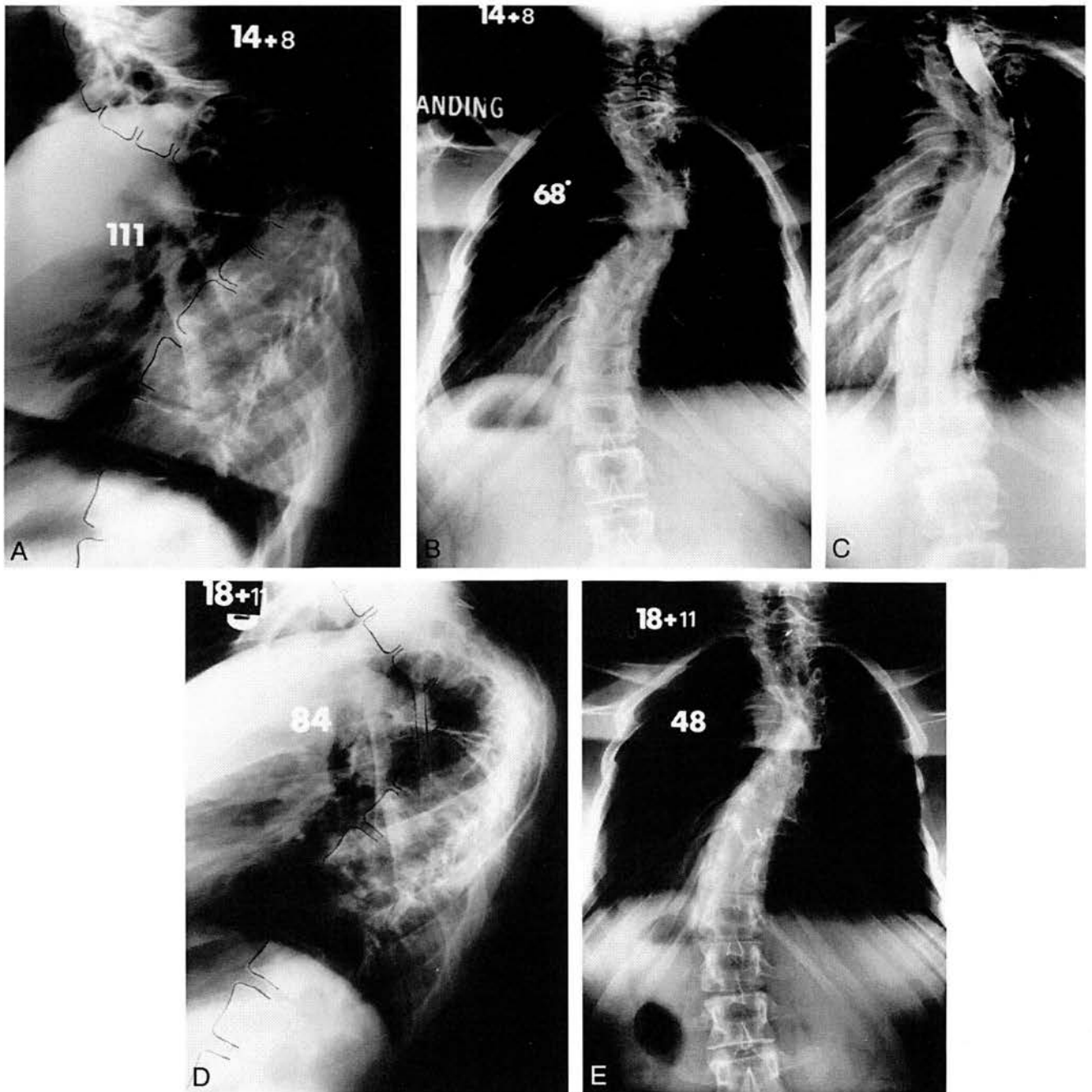


Figure 3. **A, B, C**, A boy aged 14 years 8 months with a congenital thoracic kyphoscoliosis caused by a posterolateral quadrant vertebra at T4. The kyphosis measures 111° and the scoliosis 68°. A myelogram shows spinal cord compression at the apex of the deformity, which caused a spastic paraparesis. **D, E**, The spinal cord was indirectly decompressed by partially reducing the deformity by means of an anterior release and rib strut graft correction followed by a posterior arthrodesis. The patient made a complete neurologic recovery, and at the age of 18 years 11 months there was a solid stable fusion with the kyphosis measuring 84° and the scoliosis 48°.

and the arthrodesis extended over eight vertebrae (range, 7–12). After surgery, the mean kyphosis was corrected by a mean 18° (range, 3–30°). The mean loss in correction at 1 year was 2° (range, 0–5°). No patient received additional surgery. The seven patients were observed for a mean 5 years 4 months (range, 3 years to 8 years 2 months), and all reached skeletal maturity. At this time the mean kyphosis was 78° (range, 60–98°), and no patient had lost more than 5° of correction.

Two of the three patients with a preoperative neural deficit had improvement in their neurologic status after surgery. In both patients (Patients 4 and 5), the spinal cord was indirectly decompressed by partially reducing the deformity. One patient recovered to normal (Patient 4, Figure 3), and the other (Patient 5) made a partial recovery, regaining the ability to walk unaided and with normal bladder and bowel control. The condition of the third patient (Patient 6), who underwent direct anterior

decompression of the spine cord, deteriorated neurologically, and she lost all motor power in the lower limbs but retained sensation and bowel and bladder control.

Combined Anterior and Posterior Spine Arthrodesis With Instrumentation

There were nine patients in this group. The deformity was Type I in four patients and Type III in five patients. No patient had a neurologic deficit.

The surgical treatment was similar to that the previous group except that in addition all underwent posterior instrumentation. The four patients with anterior failure of vertebral formation (Type I deformity) underwent first-stage thoracotomy and anterior spinal release by multiple disc excisions combined with fibular (1 patient) or rib (1 patient) strut grafts or vascularized rib strut graft (2 patients). The five patients with anterolateral unsegmented bar and contralateral hemivertebrae (Type III deformity) underwent osteotomy of their unsegmented bar and anterior release combined with rib strut grafts (4 patients) or a fibular strut (1 patient). After 5 days, all nine patients underwent second-stage posterior arthrodesis by use of autogenous iliac crest bone grafts combined with Harrington instrumentation in seven patients, Luque instrumentation in one patient, and AO USS in one patient. After surgery, an underarm jacket was worn for 6 months.

The mean age at surgery was 13 years 8 months (range, 9 years 7 months to 19 years), and the mean kyphosis was 90° (range, 62–126°). The mean extent of the kyphosis was over six vertebrae (range, 4–11), and the arthrodesis extended over 11 vertebrae (range, 8–14). After surgery, the kyphosis was corrected by a mean 32° (range, 14–53°).

The mean loss in correction at 1 year was 3° (range, 0–15°). The patient who lost 15° of correction had a hook displacement occurring shortly after surgery, after which the spine stabilized. No patient received additional surgery. The nine patients were observed for a mean 4 years 3 months (range, 2 years to 8 years 5 months), and all reached skeletal maturity. At this time the mean kyphosis was 59° (range, 45–82°), and only the patient with the hook displacement had lost more than 10° of correction. All had a solid stable fusion.

Comparisons Between Study Groups

Of the patients aged over 5 years, those treated by combined anterior and posterior spine surgery achieved significantly greater immediate correction ($P < 0.001$) and showed significantly less loss of correction at final follow-up ($P < 0.01$) than those receiving posterior surgery alone.

Discussion

There is no effective conservative treatment for congenital kyphosis or kyphoscoliosis.^{4,10,16} A Type I deformity caused by anterior failure of vertebral body formation has a bad prognosis, and all require immediate surgical treatment to prevent progression and possible neurologic complications caused by spinal cord compression. A Type II deformity caused by anterior failure of vertebral

body segmentation is less common and has a better prognosis with no potential for neurologic complications. Surgery in these patients is necessary only if there is already a significant deformity that requires correction or a smaller curve that shows signs of progression while the patient is under observation. A Type III deformity caused by a combination of anterior failure of vertebral body formation and segmentation is least common but progresses most rapidly, and all require immediate surgery to prevent severe deformity.¹⁰ The type of surgical treatment depends on the age of the patient, the type and size of the deformity, and the presence or absence of spinal cord compression. All patients require MRI assessment of the whole spine before surgery.

Although this is a retrospective study, it has the advantage of being one surgeon's experience (M.J.M.) with a relatively large number of patients with this uncommon congenital spine deformity treated at one institution with long-term follow-up. There have been very few previous reports of large numbers of treated patients.^{6,11,16–18}

A Type I deformity is best treated prophylactically before the age of 5 years by posterior spine arthrodesis.¹⁷ This relatively simple procedure produces a posterior spinal tether, which in the presence of continuing anterior longitudinal growth provides the opportunity for gradual correction if the initial deformity is relatively small. Type II and Type III deformities, in which there is anterior failure of segmentation of several vertebrae, have no potential for anterior growth and are therefore stabilized only by posterior fusion. In the series described here, there were 11 patients with Type I deformities under the age of 5 years (mean age, 2 years 5 months) whose mean kyphosis was 43° at the time of posterior arthrodesis. They were observed for a mean 11 years, during which time the kyphosis slowly improved by a mean of 15° in nine of the 11 patients (Figure 1). Of the two patients whose deformity progressed, one had pseudarthrosis and the other had too short a fusion, which became apparent only during the adolescent growth spurt, when additional vertebrae were added on to the deformity above and below the fused segment. It is very important that the posterior fusion extend not just to the most sagittally tilted vertebra at either end of the kyphosis but also to at least one additional vertebra above and below. Pseudarthrosis is very difficult to detect radiographically in these young patients, and the authors therefore recommend that kyphosis that progresses by more than 5°, 6 months after the arthrodesis, is an indication for surgical exploration, either to repair a pseudarthrosis or to add additional bone graft to strengthen a weak fusion that is bending. Even patients with apparently stable kyphosis after early posterior fusion should be carefully monitored radiographically to skeletal maturity because of the possibility of increasing deformity occurring during the adolescent growth spurt caused by adding on of additional vertebrae above and below the fused segment.

In an older patient, after the age of 5 years, with a Type I kyphosis greater than 50°, there is usually insufficient spinal growth time remaining, and the mechanical disadvantage resulting from the size of the deformity is usually too great to be overcome by continuing anterior growth in the presence of a posterior fusion. In these patients, the objective of surgery should be partial correction of the deformity and the creation of a spinal fusion strong enough to prevent further progression and possible neurologic complications. It is important to fuse the full extent of the kyphoscoliotic deformity, and this requires a much longer fusion than in younger children with a smaller deformity. Posterior arthrodesis with cast correction was performed on 26 patients over the age of 5 years (mean age, 12 years) whose mean kyphosis was 70°. The deformity in these patients was much greater than in the patients operated on before the age of 5 years, and the posterior grafts were therefore subject to greater distraction and tension under load, resulting in an increased likelihood of pseudarthrosis and the arthrodesis maturing to become a solid stable fusion. Thirteen patients underwent planned exploration of their posterior fusion 6 months after initial surgery, and six were found to have a pseudarthrosis that required repair. Twenty-five patients were observed to skeletal maturity, at which time 13 patients had lost more than 10° of correction, and nine of these had a kyphosis greater than it was before surgery. These poor results were not significantly improved by the addition of posterior instrumentation to the posterior arthrodesis. This study has confirmed, along with others,^{11,18} that posterior fusion alone, with or without instrumentation, is unlikely to stabilize a Type I or Type III kyphotic deformity greater than 50°. However, it may be more successful for a Type II kyphosis, in which there is already an anterior failure of segmentation, providing no correction is required.

The most successful results in patients with a kyphosis greater than 50° was achieved in those who were treated by anterior spinal release with strut graft correction and arthrodesis combined with posterior arthrodesis. This produced a significantly greater degree of correction ($P < 0.001$) and a more stable fusion ($P < 0.01$) than in the patients treated by a posterior fusion alone. Sixteen patients were treated by combined anterior and posterior surgery; their mean age at surgery was 13 years, and their mean kyphosis was 90°. The addition of posterior spine instrumentation in nine of these patients produced a better mean correction of 32°, compared with a mean 18° in those without instrumentation. Solid stable fusion developed in all, and all were observed to skeletal maturity, when their mean loss of correction was only 3°.

Unfortunately, it was not always possible to apply the adolescent-size posterior instrumentation, available at the time of this study, to these young children because of the small size of their vertebral anatomy. The posterior instrumentation was also much less effective in large rigid angular curves, especially in the upper thoracic region, and was often prominent beneath the skin over the apex of the ky-

phosis as well as obscuring the area available for posterior arthrodesis. The more modern types of downsized pediatric instrumentation are more easily applied and less bulky. However, any form of posterior instrumentation used to correct the kyphosis should be applied to the spine by use of compression and leverage but without distraction, which could result in neurologic complications.

An anterior strut graft provides the best mechanical support for a large kyphosis if it is placed far anterior to the apex of the curve and spans a long segment.¹⁴ However, a free fibular or rib strut graft placed in this isolated position has difficulty in being revascularized and may take up to 2 years to reconstitute and incorporate by a process of creeping substitution. It has been reported that such grafts, if placed more than 4 cm anterior to the apical vertebral body, are more likely to fracture during this period of incorporation, resulting in a loss of anterior support.¹ However, a vascularized rib graft that is rotated into position, maintaining its own blood supply through a vascular pedicle, has the advantage that it is living bone and does not require to be reconstituted. It heals, at either end, to the spine over a period of weeks in a manner similar to that of a fractured rib and provides lasting stability.^{2,12} In addition, the mechanical stress of supporting the spine encourages the vascularized rib to hypertrophy and provide increasing strength (Figure 2). Four patients were treated in this manner and all had successful results.

Six patients in this series presented before surgery with lower limb spastic paraparesis caused by anterior spinal cord compression at the apex of the deformity (Table 2). This complication occurred only in association with Type I deformities and was caused by posterior extrusion of a posterolateral quadrant vertebra or a posterior hemivertebra into the spinal canal at the apex of the kyphosis or kyphoscoliosis as the deformity increased. The true prevalence of this complication in untreated patients is unknown, but it has been reported to occur in up to 18% of Type I deformities^{10,16} and if left untreated always progresses to paraplegia. Laminectomy and preoperative traction are both absolutely contraindicated. Traction initially corrects the mobile compensatory curves above and below the more rigid kyphosis. This lengthens the spine and pulls the spinal cord against the unyielding apex of the kyphosis, resulting in further neurologic deterioration. Laminectomy is ineffective because the spinal cord is not compressed posteriorly, and removal of the posterior bony structures destabilizes the spine, resulting in more rapid progression of the kyphosis and greater anterior spinal cord compression.

Of the six patients with a preoperative neurologic deficit (Table 2), three were treated by direct anterior or anterolateral decompression of the spinal cord by excision of the anterior gibbus. The remaining three underwent indirect decompression by partial reduction of the deformity without direct exposure of the spinal cord. Of the three patients who underwent direct anterior decompression, one made a complete recovery, one made a partial recovery, and one was worse. Of the three pa-

tients who underwent indirect decompression by partial reduction of the deformity, two made a complete recovery, one made a partial recovery, and none was worse.

Direct anterior decompression of the spinal cord can be a hazardous procedure. The spinal cord is tightly stretched and compressed against the anterior angular gibbus, which usually lies in the middle or upper thoracic region, where the segmental blood supply to the spinal cord is poorest, the spinal canal is narrowest, and the cord is least mobile. In addition, the segmental and small vessel blood supply to the spinal cord may be congenitally abnormal or deficient in this anomalous region. Successful surgery requires excision of the anterior gibbus without inadvertent application of additional pressure on the spinal cord or impairment of its segmental blood supply. However, adequate decompression requires not only removal of the body of the anomalous apical vertebra but also partial resection of the adjacent vertebral bodies above and below. This may be difficult to achieve without interfering with the bilateral segmental blood supply to the spinal cord, which could result in ischemia and neurologic deterioration. It is therefore recommended that if there is a relatively mild neurologic deficit and the patient is still able to walk unaided, the spinal cord be indirectly decompressed. This is achieved by partial reduction of the deformity by means of an anterior spine release and strut graft correction, followed by posterior arthrodesis with instrumentation (if possible) without direct exposure of the spinal cord (Patient 4, Figure 3). However, if there is a more severe neurologic deficit with an inability to walk unaided, it is necessary, despite the risks, to perform a direct decompression by excising the anterior gibbus either through a transthoracic approach or by costotransversectomy, combined with anterior strut fusion and posterior arthrodesis with instrumentation (if possible). If the spinal cord is not adequately decompressed and the deformity is not stabilized, the patient's condition will deteriorate to paraplegia.

In conclusion, the authors recommend that all patients with Type I or Type III congenital kyphosis or kyphoscoliosis be treated at an early stage, when they are under 5 years of age, by means of posterior arthrodesis before the kyphosis exceeds 50°. A kyphosis that does not reduce to less than 50°, as measured on the lateral radiograph of the spine made with the patient supine, is best treated by an anterior release and arthrodesis with strut grafting, followed by posterior arthrodesis with instrumentation (if possible) to correct and stabilize the deformity. A Type II kyphosis does not necessarily require immediate surgical treatment unless the deformity is already sufficiently severe to require correction or there is a smaller curve shows signs of progression while the patient is under radiographic observation.

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Key Points

- Congenital kyphosis or kyphoscoliosis is an uncommon deformity, which in some patients can lead to spinal cord compression and paraplegia.
- Patients with Type I and Type III deformities are best treated before the age of 5 years by posterior arthrodesis before the kyphosis exceeds 50°.
- Larger curves require an anterior spine release and arthrodesis with strut grafting followed by posterior arthrodesis with instrumentation (if possible).

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CONGENITAL ANOMALIES OF THE RIBS AND CHEST WALL ASSOCIATED WITH CONGENITAL DEFORMITIES OF THE SPINE

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Background: Congenital anomalies of the ribs and chest wall as well as Sprengel deformity of the shoulder are often associated with congenital deformities of the spine. It has been suggested that rib anomalies may adversely affect the progression of the spinal deformity.

Methods: We conducted a retrospective study of the medical records and spine radiographs of 620 consecutive patients with congenital deformities of the spine; 497 patients (80%) had scoliosis; eighty-eight patients (14%), kyphoscoliosis; and thirty-five patients (6%), kyphosis. The rib anomalies were classified into simple and complex, and the presence of a Sprengel deformity of the shoulder was recorded. The rate of scoliosis deterioration without treatment before the age of eleven years, as well as the patient age and curve size at the time of surgery, was compared for different types of vertebral abnormalities in patients with and without rib anomalies.

Results: A total of 119 patients (19.2%) had rib anomalies, which were most commonly associated with congenital scoliosis (111 patients; 93%) and were much less frequently associated with congenital kyphoscoliosis or kyphosis (eight patients). The rib anomalies were simple in ninety-five patients and complex in twenty-four. Eighty-five patients (71%) with rib abnormalities had a scoliosis due to a unilateral failure of vertebral segmentation, and seven patients had mixed or unclassifiable vertebral anomalies. In contrast, only sixteen of 203 patients with a scoliosis due to a hemivertebra alone had rib anomalies. The rib anomalies were most frequently associated with a thoracic or thoracolumbar scoliosis (102 patients; 92%) and occurred on the concavity in eighty-two patients (74%), the convexity in twenty-two patients (20%), and were bilateral in seven patients. The Sprengel deformity occurred in forty-five patients and most frequently in association with a thoracic scoliosis due to a unilateral failure of vertebral segmentation (twenty-seven patients). No significant difference was detected in the rate of curve progression without treatment in patients with and without rib anomalies. The only exception was the mean age at the time of surgery, which was higher for patients with a unilateral unsegmented bar without rib anomalies ($p = 0.005$). In addition, no significant difference was found with regard to any tethering effect due to the site of the rib fusions on the concavity of the scoliosis, i.e., whether they were in close approximation to the spine or were more lateral ($p > 0.05$).

Conclusions: Congenital rib anomalies occur most commonly on the concavity of a thoracic or thoracolumbar congenital scoliosis that is due to a unilateral failure of vertebral segmentation, and they do not appear to have an adverse effect on curve size or rate of progression.

Level of Evidence: Prognostic Level II. See Instructions to Authors for a complete description of levels of evidence.

Congenital malformations of the ribs and chest wall, as well as anomalies of the scapulae, are often found in association with congenital deformities of the spine. These developmental chest wall abnormalities, which may be simple or complex, are due to either a failure of segmentation or formation of the ribs.

Campbell et al. stated that extensive rib fusions affecting the hemithorax on the concavity of a congenital scoliosis in growing children can act as a powerful lateral tether to further unbalance the growth of the spine, which is already being deformed by asymmetrical vertebral growth¹. In addition, an extensive thoracic congenital scoliosis associated with fused ribs

may affect thoracic function and the growth of the lungs in young children and lead to a thoracic insufficiency syndrome. An imbalance in the mechanical thrust of the ribs may also adversely affect spinal growth as well as the function of trunk muscles and the pressure within the thorax²³. Shahcheraghi and Hobbi, in a study of sixty patients with congenital scoliosis, reported that sixteen had fusion of the ribs⁴. They found that fused ribs on the concave side of a lower thoracic curve increased the rate of curve progression⁴.

In order to overcome the problem of a congenital scoliosis associated with chest wall anomalies producing thoracic insufficiency, Campbell and Hell-Vocke developed the surgical technique of expansion thoracoplasty, in which the concave hemithorax is lengthened and stabilized by serial rib distractions with use of a vertical expandable prosthetic titanium rib⁵.

To the best of our knowledge, no large study has been done on the prevalence of rib and chest wall anomalies associated with congenital deformities of the spine and the effect that these may have on curve progression. The purpose of our study was twofold: first, to document the prevalence and describe the different types of congenital rib and chest wall abnormalities as well as Sprengel deformity of the shoulder occurring in association with all types of congenital spine deformities, and, second, to assess the effect of congenital rib anomalies on the rate of progression of the spinal deformity.

Embryological Development of the Ribs, Spine, and Scapulae

The embryological development of the ribs and spine are very closely associated, and the complete anatomical pattern is formed in mesenchyme during the first six weeks of intrauterine life. Developmental abnormalities of the ribs and spine may occur during this period, and, once the mesenchymal mold is established, the cartilaginous and osseous stages follow that pattern.

Vertebral anomalies occurring during the mesenchymal stage may be due to either a unilateral defect of formation or segmentation of the primitive vertebrae and can result in a unilateral imbalance in the longitudinal growth of the spine producing a congenital scoliosis^{6,7}. Vertebral anomalies may also occur during the subsequent chondrification stage and are thought to be due to a localized failure of vascularization of the developing cartilaginous centrum. This results in varying degrees of failure of formation of the vertebral body producing a congenital kyphosis or kyphoscoliosis^{6,7}. In the late chondrification and ossification stages, osseous metaplasia may occur in the anterior part of the anulus fibrosis and ring apophysis, producing an anterior or anterolateral unsegmented bar, which can also result in a congenital kyphosis or kyphoscoliosis⁸.

The ribs form from costal processes, which are small lateral mesenchymal condensations of the developing thoracic somites and contribute cells to all parts of the developing ribs⁹⁻¹³. The distal tips of the costal processes elongate to

form ribs only in the thoracic region of the spine. Rib anomalies probably occur during the process of segmentation and resegmentation of the developing somites, after which the ribs come to articulate between the definitive thoracic vertebrae. The ribs develop into cartilaginous precursors that ossify during the fetal period.

The scapula develops embryologically along with the arm. The arm bud appears in the third week of embryonic life as a small swelling opposite the vertebral segments from the fifth cervical to the first thoracic vertebra. The scapula appears in mesenchyme during the fifth week and gradually migrates caudally. By the end of the third fetal month, the scapula reaches its final anatomical position located lateral to the spine and extending from the second to the seventh or eighth thoracic vertebra. Occasionally, the scapula may fail to fully descend to its normal location and remains in a permanently elevated position commonly known as a Sprengel deformity.

Materials and Methods

We reviewed the medical records and spine radiographs of 620 consecutive patients with a congenital spine deformity who were seen at our institution between 1960 and 2003. All patients were followed and treated by the senior author (M.J.M.), who has reported previously on the natural history and surgical management of these patients^{14,15}. There were 400 female and 220 male patients. Four hundred and ninety-seven patients (80%) had a scoliosis, eighty-eight (14%) had a kyphoscoliosis, and thirty-five (6%) had a kyphosis. Patients who had myelomeningocele, Scheuermann disease, neurofibromatosis, skeletal dysplasia, infection, or trauma were excluded.

The vertebral anomalies causing the spine deformity as well as the presence of rib anomalies or a Sprengel deformity of the shoulder were detected on anteroposterior and lateral spine and chest radiographs. We were unable to assess the prevalence of Klippel-Feil deformity because not all of our patients had cervical radiographs. The vertebral anomalies and the type of spine deformity were categorized according to the classification proposed by Winter et al.^{16,17} and McMaster et al.^{14,15} into failures of vertebral segmentation, failures of vertebral formation, and mixed anomalies producing a congenital scoliosis, kyphoscoliosis, or kyphosis (Tables I and II).

The site of the curvature was defined according to the classification proposed by the Scoliosis Research Society¹⁸: cervicothoracic (apex at C7 or T1), thoracic (apex between T2 and T11), thoracolumbar (apex at T12 or L1), lumbar (apex between L2 and L4), and lumbosacral (apex at L5 or caudad). There is a recognized difficulty in accurately measuring spine radiographs of patients with congenital scoliosis¹⁹. However, all of the serial spine radiographs for each patient in this study were measured by the senior author in both the coronal and sagittal planes with use of the Cobb method²⁰ with the aim of eliminating interobserver measurement error. All spine radiographs were made with the patient

TABLE I Summary of the Data on the Patients with Congenital Scoliosis

	Defects of Segmentation			Defects of Formation		Mixed or Unclassifiable Anomalies
	Unilateral Unseg- mented Bar with Contralateral Hemivertebrae	Unilateral Unsegmented Bar	Block Vertebrae	Hemivertebrae	Wedge Vertebrae	
Total no. of patients (F/M)	66 (46/20)	160 (118/42)	6 (2/4)	203 (127/76)	10 (6/4)	52 (34/18)
No. of patients with rib anomalies (F/M)	34 (24/10)	51 (38/13)	1 (0/1)	16 (12/4)	2 (2/0)	7 (4/3)
Simple	25 (18/7)	42 (30/12)	1 (0/1)	13 (10/3)	2 (2/0)	6 (4/2)
Complex	9 (6/3)	9 (8/1)	—	3 (2/1)	—	1 (0/1)
Type of curve*						
Cervical	—	—	—	0/1	—	—
Cervicothoracic	—	—	—	1/5	—	0/3
Thoracic	24/42 (57)	46/134 (34)	1/4	11/82 (13)	2/7	4/31 (13)
Thoracolumbar	10/19	4/15	0/1	0/14	0/1	0/4
Lumbar	0/5	1/9	—	1/45 (2)	0/2	0/4
Lumbosacral	—	—	—	0/15	—	—
Multiple	—	0/2	0/1	3/37 (8)	—	3/10
Side of rib anomaly†						
Concave						
Simple	25 (78)	33 (89)	1	6	1	4
Complex	7 (22)	4 (11)	—	—	—	1
Convex						
Simple	—	8	—	4	—	1
Complex	2	4	—	3	—	—
Bilateral						
Simple	—	1	—	3	1	1
Complex	—	1	—	—	—	—
Sprengel deformity (no. of patients)	6	21	1	3	3	9
Concave	5	12	—	1	3	5
Convex	1	9	1	2	—	4

*The values are given as the number of patients with rib anomalies/total number of patients with curve, with the percentage in parentheses.

†The values are given as the number of patients, with the percentage of the total for that side of the curve for each group given in parentheses.

standing, except for patients in early infancy for whom radiographs were made in the supine position. Care was taken to measure all curves with use of the same anatomical landmarks on the serial spine radiographs.

We classified the rib anomalies seen in our patients as *simple* or *complex*. A patient with a *simple* rib anomaly had only one of the following: a localized fusion of two or three ribs (Fig. 1) or a small chest wall defect that was due to a deviation of one or two ribs or an absence of a rib. Two patients had a bifid rib, which is a single rib separating into two ribs more distally, and one patient had a duplication of two ribs arising from the same vertebral segment. A patient with *complex* rib anomalies had multiple extensive rib fusions, usually without a set pattern, combined with an adjacent large chest wall defect that was due to an absence or deviation of ribs (Figs. 2, 3, and 4). Two patients had a bifid rib combined with

either a large chest wall defect (Fig. 5) or multiple fused ribs. The location of the rib anomalies in relation to the convexity or concavity, as well as the apex of the spine deformity, was recorded. The radiographs in Figures 1 through 5 were reproduced with use of photographic contrast imaging in order to show the rib and shoulder anomalies.

The presence of an abnormal number of normally formed ribs was not considered to be an anomaly for the purpose of this study. Sixty-three (10%) of the 620 patients had an abnormal number of ribs, ranging from seven to fifteen, either unilaterally or bilaterally. An abnormal number of ribs was seen most often in patients who had a scoliosis due to a unilateral failure of vertebral segmentation (forty-five patients; 71%) ($p < 0.01$).

In order to assess the possible effect of the rib anomalies, either simple or complex, on the congenital scoliosis, we esti-

TABLE II Summary of the Data on the Patients with Congenital Kyphosis or Kyphoscoliosis

	Defects of Segmentation	Defects of Formation			Mixed Anomalies	Unclassifiable Anomalies
	Anterior or Anterolateral Unsegmented Bar	Posterolateral Quadrant or Posterior Hemivertebrae	Wedge Vertebrae	Butterfly Vertebrae	Anterolateral Bar and Contralateral Quadrant Vertebrae	
Total no. of patients (F/M)	26 (16/10)	51 (31/20)	9 (1/8)	15 (9/6)	12 (6/6)	10 (4/6)
No. of patients with rib anomalies (F/M)	2 (2/0)	1 (0/1)	—	2 (1/1)	2 (1/1)	1 (1/0)
Simple	1 (1/0)	1 (0/1)	—	1 (0/1)	2 (1/1)	1 (1/0)
Complex	1 (1/0)	—	—	1 (1/0)	—	—
Type of curve (no. of patients with rib anomalies/total no. with curve)						
Thoracic	1/12	0/29	0/5	1/10	0/2	0/7
Thoracolumbar	1/9	1/20	0/4	1/5	2/8	1/2
Lumbar	0/5	0/2	—	—	0/2	0/1
Side of rib anomaly (no. of patients)						
Concave						
Simple	1	—	—	—	1	1
Complex	—	—	—	—	—	—
Convex						
Simple	—	—	—	—	—	—
Complex	—	—	—	—	—	—
Bilateral						
Simple	—	—	—	—	—	—
Complex	1	—	—	—	—	—
Associated with kyphosis						
Simple	—	1	—	1	1	—
Complex	—	—	—	1	—	—
Sprengel deformity (no. of patients)						
Concave	—	1	—	—	—	—
Convex	—	—	—	—	—	—
Associated with kyphosis	—	—	—	1	—	—

rated the rate of progression of the untreated curves before the age of eleven years, as well as the patient age and size of the scoliosis before spine surgery and compared these findings with those in patients with similar spine deformities but without rib anomalies. We also evaluated the possible tethering effect due to the site of the rib anomalies on the concavity of the scoliosis, i.e., whether the rib anomalies were in close association to the spine or were more lateral.

Statistical Methods

The prevalences of congenital rib anomalies among patients with different types of vertebral anomalies causing a scoliosis, kyphoscoliosis, or kyphosis were compared with use of the chi-square test (with continuity correction). Due to the low preva-

lence of rib anomalies in patients with congenital kyphosis, the analysis was performed with use of the Fisher exact test.

Analysis of variance was used to compare the yearly rate of scoliosis progression without treatment, as well as the age and curve size at the time of spine surgery, among patients with or without rib anomalies, either simple or complex. When significant differences were found, the t test was used to examine this association in more detail by grouping the three rib abnormality groups into two (those with or without rib anomalies). The t test was also used to compare the tethering effect produced by concave rib fusions either in close proximity to or more lateral and separate to a unilateral failure of vertebral segmentation. P values of <0.05 were considered to be of significance for all statistical tests.



Fig. 1

Radiograph of a patient with congenital scoliosis due to a unilateral unsegmented bar. There is a simple rib anomaly with fusion of two ribs on the concavity of the curve (white arrow).

Results

The type and distribution of the rib anomalies (simple and complex) and the Sprengel deformity of the shoulder in patients with congenital scoliosis, kyphoscoliosis, and kyphosis due to different types of vertebral abnormalities, as well as the type and location of the spinal curves, are shown in Tables I and II.

Rib anomalies occurred in 119 (19.2%) of the 620 patients with congenital deformities of the spine. The female-to-male ratio was 2.5:1 for the patients with rib anomalies compared with 1.7:1 for the patients without rib anomalies.

Type of Spine Deformity

Congenital Scoliosis

Rib anomalies occurred in 111 (22.3%) of the 497 patients with congenital scoliosis (Table I). These rib anomalies were most frequently associated with a unilateral failure of vertebral segmentation ($p < 0.001$). Of the 160 patients with a unilateral unsegmented bar alone, fifty-one (32%) had rib anomalies. Of the sixty-six patients with a unilateral unsegmented bar combined with contralateral hemivertebrae at the same level,

thirty-four (52%) had rib anomalies. In contrast, patients with a unilateral failure of vertebral formation had a low prevalence of rib anomalies. Of the 203 patients with hemivertebrae alone, only sixteen had rib anomalies.

Congenital Kyphoscoliosis and Kyphosis

Rib anomalies occurred in only eight of the 123 patients with a congenital kyphoscoliosis or kyphosis (Table II). No significant difference was detected in the distribution of rib anomalies between patients with a unilateral failure of vertebral formation and those with a failure of segmentation ($p = 0.897$).

Site of Spine Deformity

Rib anomalies were most frequently associated with thoracic or thoracolumbar curves.

Congenital Scoliosis

Of the 300 patients with a thoracic scoliosis, eighty-eight (29%) had rib anomalies. Of the fifty-four patients with a thoracolumbar scoliosis, fourteen had rib anomalies. In contrast, rib anomalies occurred in only two of the eighty patients with

a lumbar or lumbosacral scoliosis ($p < 0.001$) (Table I).

Congenital Kyphoscoliosis and Kyphosis

Rib anomalies were infrequent in this group. Of the sixty-five patients with a thoracic curve, only two had rib anomalies. Of the forty-eight patients with a thoracolumbar curve, six had rib anomalies. None of the ten patients with a lumbar curve had rib anomalies ($p < 0.001$) (Table II).

Type of Rib Anomalies

Of the 119 patients with rib anomalies, ninety-five (80%) had

simple anomalies and twenty-four (20%) had complex anomalies. The most common simple rib anomaly, occurring in seventy-two patients (76%), was a localized fusion of two or three ribs (Fig. 1). The most common complex rib anomaly, occurring in twenty-one patients (88%), was an extensive fusion of multiple ribs associated with a large adjacent chest wall defect (Fig. 3).

Congenital Scoliosis

Simple rib anomalies occurred in eighty-nine (18%) of the 497 patients with congenital scoliosis, and sixty-eight (76%)

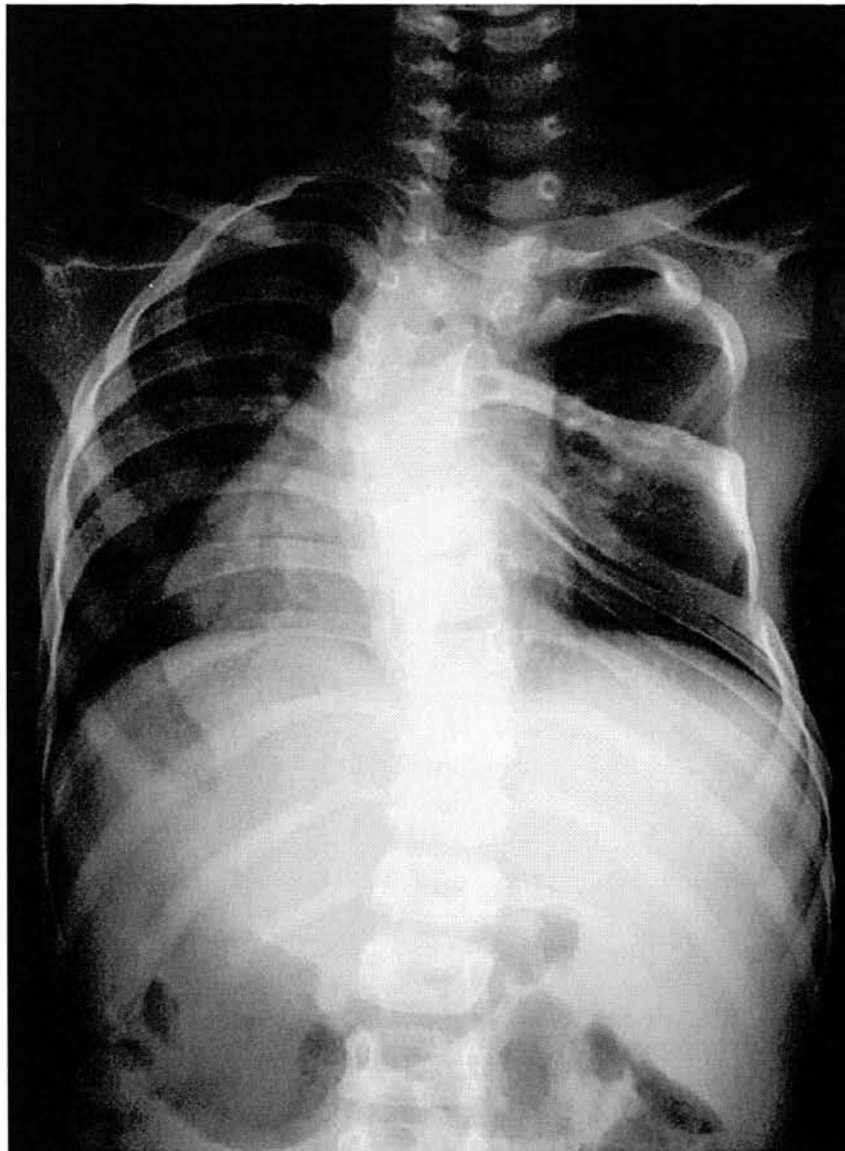


Fig. 2

Radiograph of a patient with congenital scoliosis due to a unilateral unsegmented bar. There are two fused ribs and two chest wall defects on the concavity of the curve (a complex anomaly).

of the eighty-nine patients had a localized fusion of two or three ribs (Fig. 1). Complex rib anomalies occurred in twenty-two patients (4.4%), and all had a combination of fused ribs with chest wall defects (Figs. 2, 3, and 4).

Congenital Kyphoscoliosis and Kyphosis

Simple rib anomalies occurred in only six patients with con-

genital kyphoscoliosis or kyphosis; four patients had a fusion of ribs. Complex rib anomalies occurred in only two patients. One patient with a complex rib anomaly had a chest wall defect with an ipsilateral bifid rib, and another had a combination of fused ribs with a contralateral bifid rib. Complex rib anomalies also occurred in a patient with a thoracolumbar kyphosis.

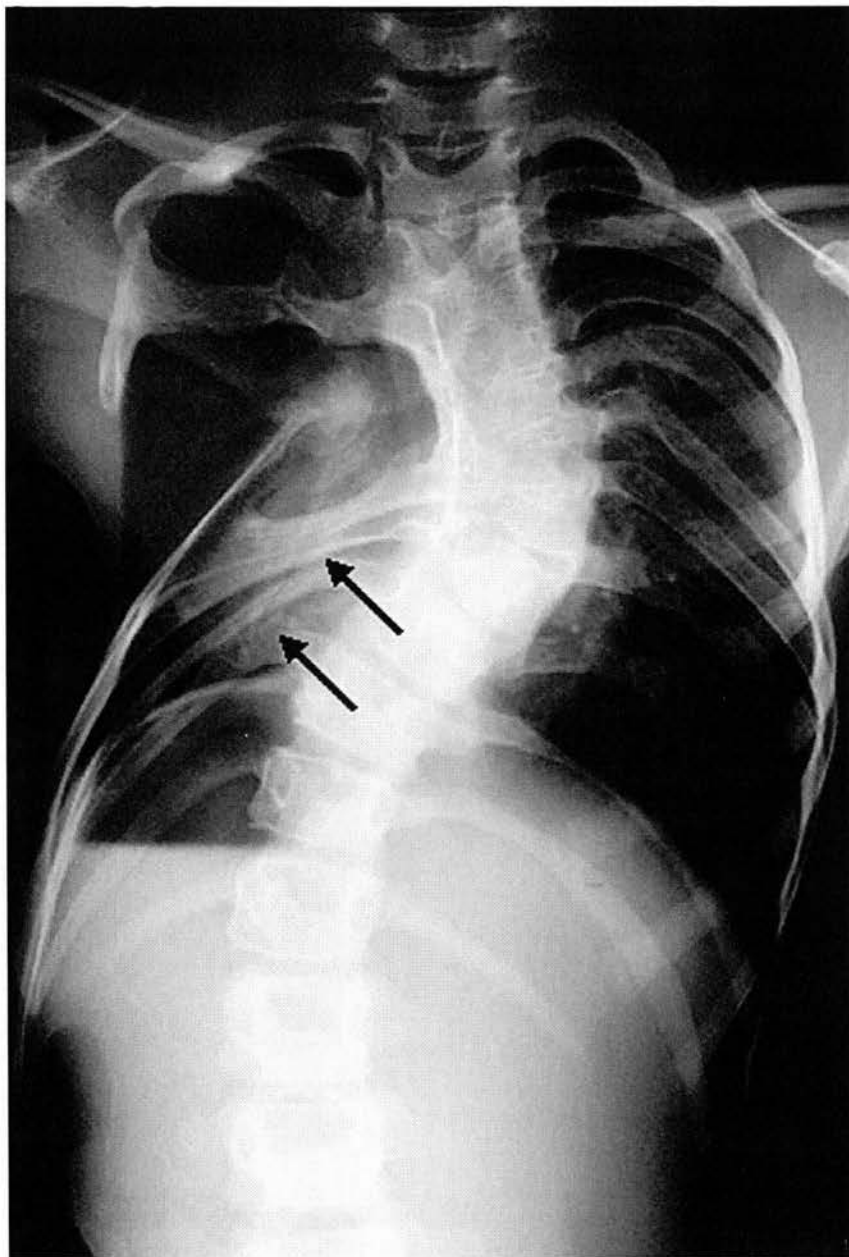


Fig. 3

Radiograph of a patient with congenital scoliosis due to a unilateral unsegmented bar. There are complex rib anomalies on the concavity of the curve with fusion of two pairs of ribs (black arrows) and a large thoracic wall defect as well as congenital elevation of the scapula on the concave side.

Side and Site of Rib Anomalies in Relation to the Scoliosis

Simple rib anomalies occurred most frequently on the concavity of the scoliosis (seventy [79%] of eighty-nine patients) and were less common on the convexity (thirteen patients). Six patients had simple rib anomalies bilaterally. Complex rib anomalies occurred on the concavity of the scoliosis in twelve patients and on the convexity in nine patients. One patient had complex rib anomalies bilaterally.

Rib anomalies, both simple and complex, occurring on the concavity of the scoliosis were most frequent in patients with a unilateral failure of vertebral segmentation (sixty-nine patients; $p < 0.001$). These anomalies were present in thirty-seven (23%) of the 160 patients with a unilateral unsegmented bar alone and in thirty-two (48%) of the sixty-six patients with a unilateral unsegmented bar with a contralateral hemivertebra at the same level. Twenty-three of the patients with a unilateral

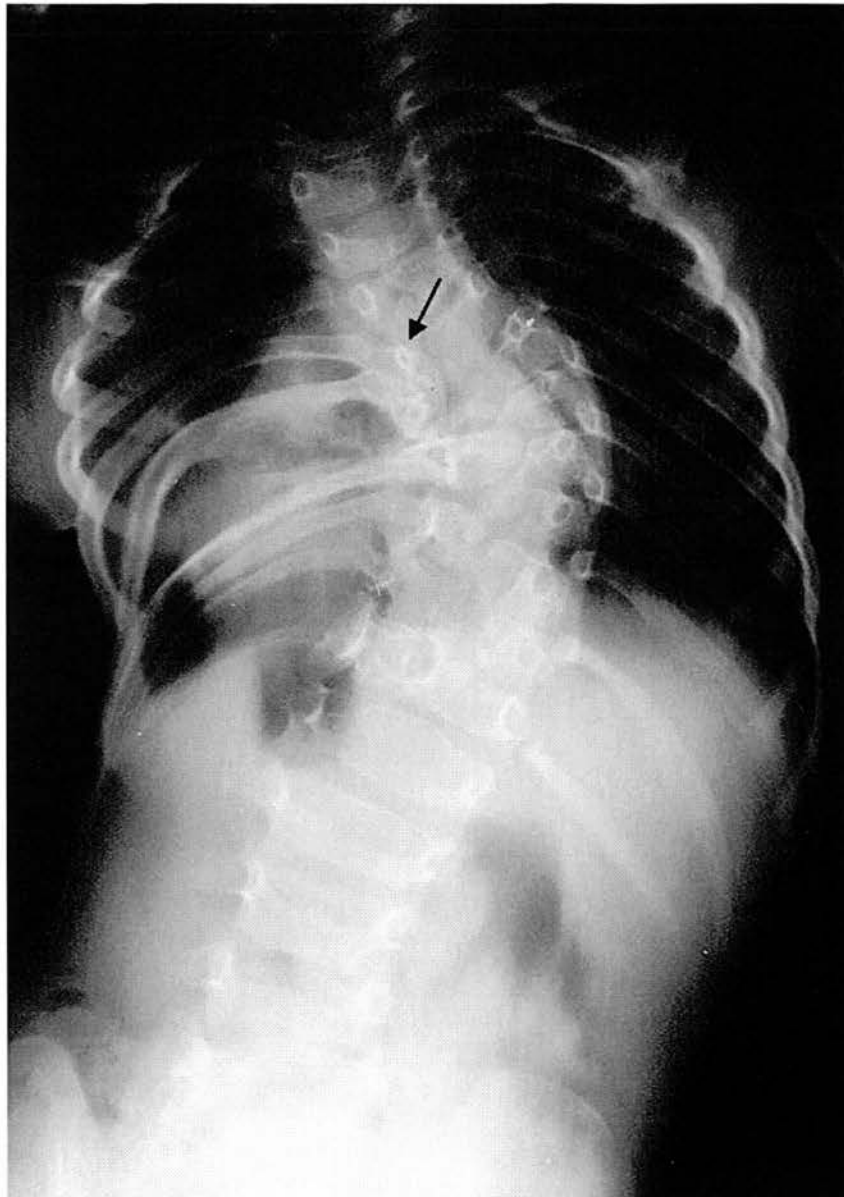


Fig. 4

Radiograph of a patient with congenital scoliosis due to a unilateral unsegmented bar with contralateral hemivertebrae. There are complex rib anomalies on the concavity of the curve with the fusion of two ribs adjacent to the osseous bar (black arrow) and a thoracic wall defect. There is evidence of a diastematomyelia osseous spur at the thoracolumbar junction.

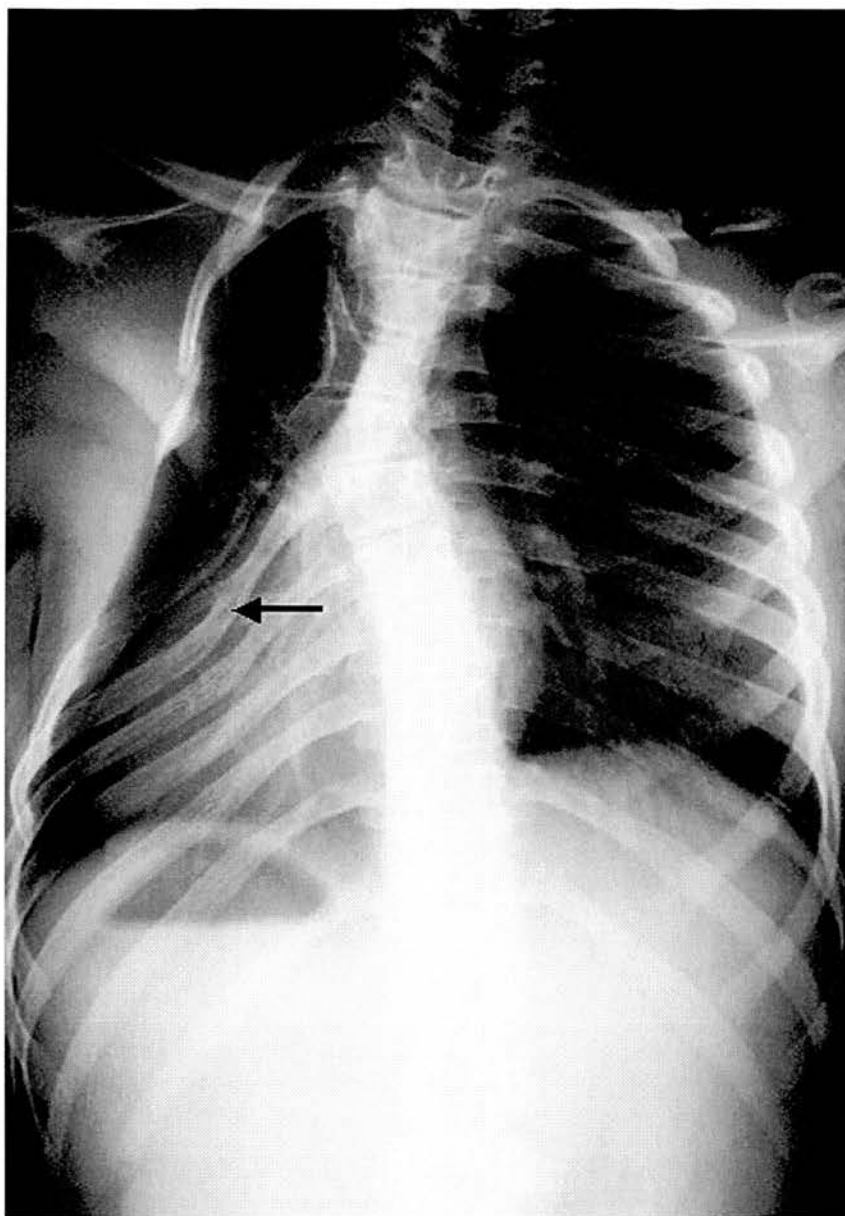


Fig. 5

Radiograph of a patient with congenital scoliosis due to a unilateral unsegmented bar. There are complex rib anomalies with a bifid rib (black arrow) and an ipsilateral large chest wall defect, as well as a Sprengel deformity on the convexity of the curve.

failure of vertebral segmentation had rib fusions on the concavity of the scoliosis, which were in close association with the unsegmented bar. Twenty of them had simple rib anomalies, and three had complex rib anomalies. An additional thirty-eight patients had concave rib fusions, but these lay more lateral and separate from the spine. Thirty of them had simple rib anomalies, and eight had complex rib anomalies.

Rib anomalies on the convexity of the scoliosis also oc-

curred most frequently in patients with a unilateral failure of vertebral segmentation (fourteen patients) and all anomalies were separate from the spine.

Rate of Scoliosis Progression in Untreated Patients with and without Rib and Chest Wall Anomalies

Of the 119 patients who had simple or complex rib abnormalities, the majority (eighty-five patients; 71%) had anomalies

that occurred on the concavity of a thoracic or thoracolumbar scoliosis due to either a unilateral unsegmented bar or a unilateral unsegmented bar with contralateral hemivertebrae at the same level. We compared the rate of curve progression per year before the age of eleven years in the untreated patients with and without rib anomalies who had been followed for more than two years (mean, six years; range, two to 10.4 years) (Table III).

For the patients with other types of spine deformities, the number of those with rib anomalies was insufficient to make a meaningful comparison.

Patients with a Unilateral Unsegmented Bar and Contralateral Hemivertebrae at the Same Level

Twenty-two patients with simple rib anomalies were followed for a mean of five years (range, two to 10.2 years) and had a mean rate of curve progression of 6.2° per year. Six patients with complex rib abnormalities were followed for a mean of 5.6 years (range, two to nine years) and had a mean rate of scoliosis deterioration of 5.7° per year. Thirteen patients without rib anomalies were followed for a mean of seven years (range, 2.4 to 10.4 years) and had a mean rate of curvature progression of 5.6° per year.

No significant difference was detected in the rate of scoliosis progression per year for patients with simple rib anomalies, with complex rib anomalies, or without rib anomalies ($p = 0.85$) (Table III).

Patients with a Unilateral Unsegmented Bar Alone

Twenty-one patients with simple rib anomalies were followed for a mean of 4.8 years (range, two to 9.9 years) and had a mean rate of curve progression of 2.8° per year. Five patients with complex rib anomalies were followed for a mean of 5.4 years (range, two to 9.1 years) and had a mean rate of scoliosis progression of 4.4° per year. Twenty-eight patients without rib anomalies were followed for a mean of six years (range, two to 10.4 years) and had a mean rate of curvature progression of 3.9° per year.

No significant difference was detected in the rate of

scoliosis progression per year among patients with simple rib anomalies, with complex rib anomalies, or without rib anomalies ($p = 0.29$) (Table III).

Age and Severity of Scoliosis Before Spinal Arthrodesis in Patients with and without Rib and Chest Wall Anomalies

We estimated the age and the size of the scoliotic curve at the time of spinal arthrodesis in the patients with and without concave rib abnormalities who had a thoracic or thoracolumbar scoliosis due to a unilateral unsegmented bar with or without contralateral hemivertebrae at the same level (Table IV).

Patients with a Unilateral Unsegmented Bar and Contralateral Hemivertebrae at the Same Level

In the twenty-three patients with simple rib anomalies, the mean age at the time of spinal arthrodesis was 8.5 years (range, 1.1 to eighteen years) and the mean curvature was 76° (range, 36° to 120°). The five patients with complex rib abnormalities underwent spinal arthrodesis at a mean age of seven years (range, 0.6 to thirteen years) and had a mean scoliosis of 62° (range, 37° to 88°). In the twenty-one patients without rib anomalies, spinal arthrodesis was performed at a mean age of nine years (range, 1.6 to sixteen years) and the mean curvature was 79° (range, 33° to 142°).

No significant difference was detected among patients with simple rib anomalies, with complex rib anomalies, or without rib anomalies with respect to the age or the curve at the time of surgery ($p = 0.64$ and 0.44 , respectively) (Table IV).

Patients with a Unilateral Unsegmented Bar Alone

In the twenty-two patients with simple rib anomalies, the mean age at the time of spinal arthrodesis was ten years (range, 1.5 to nineteen years) and the mean scoliosis was 61° (range, 33° to 100°). The ten patients with complex rib anomalies underwent spinal arthrodesis at a mean age of 11.5 years (range, 5.7 to 15.5 years) and the mean curvature was 68° (range, 54° to 95°). In the fifty-seven patients without rib anomalies, spinal

TABLE III Comparison of the Mean Rate of Curve Progression for Patients Followed Without Treatment or Before Spine Surgery

	Thoracic and Thoracolumbar Scoliotic Curves without Treatment or Before Spine Surgery									Analysis of Variance (P Value)
	Simple Rib Anomalies			Complex Rib Anomalies			Without Rib Anomalies			
	Total No. of Patients	Rate of Curve Progression* (deg/yr)	No. of Patients Who Had an Operation	Total No. of Patients	Rate of Curve Progression* (deg/yr)	No. of Patients Who Had an Operation	Total No. of Patients	Rate of Curve Progression* (deg/yr)	No. of Patients Who Had an Operation	
Unilateral unsegmented bar with contralateral hemivertebrae	22	6.2 ± 3.3	19	6	5.7 ± 3.1	4	13	5.6 ± 2.8	13	0.85
Unilateral unsegmented bar	21	2.8 ± 2.2	15	5	4.4 ± 1.3	5	28	3.9 ± 3	20	0.29
*The values are given as the mean and the standard deviation.										

*The values are given as the mean and the standard deviation.

TABLE IV Comparison of the Mean Age and Curve Size at the Time of Spine Surgery

	Thoracic and Thoracolumbar Scoliotic Curves at the Time of Surgery									Analysis of Variance (P Value)	
	Simple Rib Anomalies			Complex Rib Anomalies			Without Rib Anomalies				
	No. of Patients	Age at Time of Surgery* (yr)	Curve Size at Time of Surgery* (deg)	No. of Patients	Age at Time of Surgery* (yr)	Curve Size at Time of Surgery* (deg)	No. of Patients	Age at Time of Surgery* (yr)	Curve Size at Time of Surgery* (deg)	Age at Time of Surgery	Curve Size at Time of Surgery
Unilateral unsegmented bar with contralateral hemivertebrae	23	8.5 ± 4.1	76 ± 25.4	5	7 ± 4.5	62 ± 19.4	21	9 ± 4.8	79 ± 28	0.64	0.44
Unilateral unsegmented bar	22	10 ± 4.1	61 ± 21.7	10	11.5 ± 2.8	68 ± 12.4	57	13 ± 4.3	61 ± 19.2	0.005	0.58
*The values are given as the mean and the standard deviation.											

*The values are given as the mean and the standard deviation.

arthrodesis was performed at a mean age of thirteen years (range, 5.9 to thirty-seven years) and the mean curvature was 61° (range, 37° to 115°).

No significant difference was detected in the curve size at the time of surgery in patients with simple rib anomalies, with complex rib anomalies, or without rib anomalies ($p = 0.58$) (Table IV). However, the age at the time of surgery was significantly different among the three rib anomaly groups ($p = 0.005$). When grouping the patients into those with and those without rib anomalies, we found that the mean age at the time of surgery was higher for those without rib anomalies (mean difference, 2.84 years; 95% confidence interval, 1.03 to 4.65 years) ($p = 0.002$).

Effect of the Site of Concave Rib Fusions on the Rate of Curve Progression, Age, and Curve Size at the Time of Spinal Arthrodesis

We compared the rate of curve progression per year before the age of eleven years in patients with a thoracic or thoracolumbar scoliosis due to a unilateral failure of segmentation (with or without contralateral hemivertebrae at the same level) who had concave rib fusions located either in close association with

or more lateral and separate from the unilateral unsegmented bar and were followed without treatment for more than two years (mean, 4.7 years; range, two to 9.2 years) (Table V). We also compared the two groups with respect to the age and curve size at the time of surgery.

Patients with Rib Fusions in Close Association to the Unilateral Unsegmented Bar

The eight patients in this group were followed for a mean of 4.8 years (range, two to 9.2 years) and had a mean rate of curve progression of 4.6° per year. The mean age at the time of spinal surgery for this group of patients was 11.2 years (range, 1.1 to nineteen years) and the mean curvature was 68.2° (range, 33° to 105°). Seven patients had simple rib anomalies, and one patient had complex rib anomalies. The patient with complex rib anomalies had a combination of fused ribs with an adjacent chest wall defect.

Patients with Rib Fusions Lying Lateral and Separate from the Unilateral Unsegmented Bar

The twenty-four patients in this group were followed for a mean of 4.7 years (range, two to nine years) and had a mean

TABLE V Comparison of the Mean Rate of Curve Progression Before Surgery and the Mean Age and Curve Size at the Time of Spine Surgery

Thoracic and Thoracolumbar Scoliotic Curves Due to a Unilateral Unsegmented Bar with or without Contralateral Hemivertebrae at the Same Level				
	No. of Patients	Rate of Curve Progression* (deg/yr)	Age at Time of Surgery* (yr)	Curve Size at Time of Surgery* (deg)
Rib anomalies in close association to a unilateral unsegmented bar	8	4.6 ± 3.4	11.2 ± 5.3	68.2 ± 25.6
Rib anomalies lying lateral and separate from a unilateral unsegmented bar	24	5.4 ± 2.8	8.4 ± 3.7	73.7 ± 24.2
T test (p value)	—	0.54	0.07	0.54
Difference in mean values (95% confidence interval)	—	0.8 (−1.6 to 3.1)	2.8 (−5.3 to 0.4)	5.5 (−12.2 to 21.3)

*The values are given as the mean and the standard deviation.

rate of curve progression of 5.4° per year. The mean age at the time of spinal arthrodesis for this group was 8.4 years (range, 0.6 to 14.4 years), and the mean curvature was 73.7° (range, 33° to 120°). Eighteen patients had simple rib anomalies, and six patients had complex rib anomalies. All of the patients with complex rib anomalies had a combination of fused ribs with an adjacent chest wall defect.

With the numbers available, no significant difference was detected among patients with rib fusions in continuity with or lateral and separate to a unilateral failure of vertebral segmentation with respect to the rate of scoliosis progression per year ($p = 0.54$), the age of the patient at the time of surgery ($p = 0.07$), and the curve size at the time of surgery ($p = 0.54$) (Table V).

Sprengel Deformity

A congenital elevation of the scapula was present in forty-five (7%) of the 620 patients with congenital spine deformities. It occurred most frequently in association with a cervicothoracic or thoracic scoliosis (forty-three patients; 96%) ($p < 0.001$). Of the remaining two patients, one had a congenital kyphoscoliosis and the other had a kyphosis.

The associated scoliosis in forty-three patients was due to a unilateral unsegmented bar (with or without contralateral hemivertebrae at the same level) in twenty-seven patients (63%) and to mixed or unclassifiable vertebral anomalies in nine patients. The site of the elevated shoulder was on the concavity of the scoliosis in twenty-six patients (60%) and on the convexity in seventeen patients (40%). None of the patients included in this study had bilateral Sprengel deformity.

Rib anomalies (fourteen simple and ten complex) were also present on the same side of the elevated scapula in twenty-four (53%) of forty-five patients. In sixteen of those patients, the scoliosis was due to a unilateral unsegmented bar alone ($p < 0.001$).

Discussion

Congenital deformities of the spine are a gradually blending spectrum of deformities, ranging from a congenital scoliosis through kyphoscoliosis to a pure kyphosis. These deformities are due to an asymmetrical failure of development of one or more vertebrae resulting in a localized imbalance in the longitudinal growth of the spine and an increasing spinal curvature, which continues to progress until skeletal maturity¹⁴⁻¹⁷. The ribs are formed in close association with the vertebrae, and it is, therefore, not surprising to have a combination of developmental abnormalities affecting both the ribs and the vertebrae⁹⁻¹³.

Shahcheraghi and Hobbi reported the curve patterns and progression seen in sixty patients with congenital scoliosis⁴. Sixteen of their patients had fusion of the ribs: eight had fusion on the concavity; three, on the convexity; and five, bilaterally. No distinction was made between simple rib anomalies, complex rib anomalies, and chest wall anomalies. The spine deformity in seven of these patients was due to multiple unclassifiable vertebral abnormalities. They found that the

presence of fused ribs was associated with higher curve progression for all types of congenital vertebral anomalies except unilateral unsegmented bars with contralateral hemivertebrae⁴. However, this finding was not significant. The fusion of ribs on the concavity of a lower thoracic curvature was also associated with increased curve size and rate of progression⁴.

Other studies have been confined to two rare hereditary conditions affecting the thoracic cage and associated with congenital spinal anomalies, in which most of the vertebrae in the thoracic and thoracolumbar regions are malformed²¹⁻²⁵. Spondylothoracic dysplasia (Jarcho-Levin syndrome) is transmitted with both autosomal and recessive inheritance and consists of multiple failures of vertebral segmentation, multiple posterior rib fusions, and frequent absence of segments. Constriction of the chest and consequent respiratory failure often results in premature death in early infancy²¹⁻²⁴. Normal life expectancy is anticipated for patients with spondylocostal dysostosis, which is due to a lesser degree of rib deformation. However, marked stunting of the trunk occurs in these patients and is the result of the multiple vertebral anomalies^{24,25}. In our series, we identified only one patient with a Jarcho-Levin syndrome who died shortly after birth from respiratory failure and one patient with spondylocostal dysostosis whose spine was stunted but not otherwise deformed.

To the best of our knowledge, there has been no large study of the association between congenital spine deformities and congenital anomalies of the ribs and chest wall and the effect that these might have on curve progression. Knowledge of the natural history of congenital spine deformities with and without rib abnormalities is essential in planning prophylactic treatment for these patients at an early stage.

In our study of 620 consecutive patients with congenital spine deformities due to developmental vertebral anomalies, we classified the associated rib and chest wall anomalies according to whether they were simple or complex and investigated their potential effect on curve progression. A total of 497 patients (80%) had congenital scoliosis, eighty-eight (14%) had kyphoscoliosis, and thirty-five (6%) had kyphosis. The overall prevalence of rib anomalies was 19.2% (119 patients). However, the prevalence of rib abnormalities was the highest for patients with congenital scoliosis (111 patients; 22.3%) ($p < 0.001$) and was rare in patients with kyphoscoliosis (four patients) and kyphosis (four patients). A congenital elevation of the scapula (a Sprengel deformity) also occurred in forty-five patients and was most commonly associated with congenital scoliosis.

A possible explanation for this variation in the prevalence of rib and chest wall anomalies among the different types of congenital spine deformities is that the vertebral anomalies responsible for the development of a scoliosis appear during the mesenchymal period (the first six weeks of intrauterine life), when the ribs and scapulae are also developing. However, the vertebral anomalies responsible for a congenital kyphoscoliosis or kyphosis are believed to occur at a later stage during the chondrification and ossification periods, when the basic anatomy of the osseous structures form-

ing the ribs and scapulae has already been established^{6,7,17}.

Simple rib anomalies occurred in ninety-five patients (80%), and complex rib anomalies occurred in twenty-four patients (20%). The most common type of simple rib deformity was a localized fusion of two or three consecutive ribs (seventy-two patients; 76%) (Fig. 1), and the most frequent type of complex rib abnormality was a combination of fused ribs with a large adjacent thoracic wall defect (twenty-one patients; 88%) (Figs. 2, 3, and 4).

In this series, rib anomalies were most frequently found in patients with a congenital thoracic or thoracolumbar scoliosis due to a unilateral failure of vertebral segmentation (eighty-four [40%] of 210 patients), and they occurred most commonly on the concavity of the scoliosis (sixty-nine patients; 82%). A likely explanation for this close association between the side of the rib abnormalities and this type of congenital scoliosis is a localized unilateral embryological error resulting in a failure of segmentation of both the primitive ribs and vertebrae on the same side occurring at the same time.

Ninety-five patients with a thoracic or thoracolumbar congenital scoliosis due to a unilateral failure of vertebral segmentation with and without rib anomalies were followed without treatment for more than two years before the age of eleven years (Tables III and IV). Seventy-six (80%) of them subsequently underwent surgical treatment to control the rapidly progressive spine deformity (Table III). We found no significant difference in the rate of curve progression without treatment or the Cobb angle at the time of surgery for these patients with or without rib anomalies, either simple or complex ($p > 0.05$) (Tables III and IV). The only difference was that patients with rib anomalies associated with a unilateral unsegmented bar alone had surgical treatment at a younger age compared with those without rib anomalies ($p = 0.005$) (Table IV). We have no good explanation for this finding. A limitation of the study was the relatively small number of patients after they were subcategorized into the three rib anomaly groups (those with simple, complex, or no rib anomalies).

Theoretically, it is possible that rib fusions lying on the concavity of a scoliosis in close association with the spine may have a different tethering effect compared with similar anomalies located more laterally. In order to assess this effect, we reviewed the cases of thirty-two patients with concave rib fusions associated with a thoracic or thoracolumbar scoliosis due to a unilateral failure of vertebral segmentation who were followed without treatment for more than two years (Table V). Eight of these patients had rib fusions in close association with the unsegmented bar, and twenty-four patients had fused ribs lying more laterally. All of these patients subsequently required surgical treatment to control the rapidly progressive scoliotic deformity. We found no significant difference in the rate of curve progression without treatment or the age and Cobb angle at the time of spine surgery for these patients regardless of the location of the concave rib fusions ($p > 0.05$). A limitation of this study is the small number of patients for comparison in the two rib-fusion groups.

In our opinion, it is likely that rib fusions on the concavity of a scoliosis can cause a lateral tether and contribute to the development of the curvature. However, these rib anomalies usually occur in association with a unilateral failure of vertebral segmentation, which is recognized to produce a severe unilateral imbalance in the longitudinal growth of the spine and a rapidly progressive scoliosis. We believe that the main driving force for the development of the scoliosis in these patients is the unilateral failure of vertebral segmentation, and this greatly exceeds any adverse effect from the rib anomaly. In these circumstances, our finding that there is no apparent difference in the rate of curve progression or the size of the curve at the time of spinal arthrodesis in the patients with or without concave rib fusions regardless of their location is understandable.

Congenital elevation of the scapula (Sprengel deformity of the shoulder) occurred most commonly in association with a cervicothoracic or thoracic scoliotic deformity due to a unilateral failure of vertebral segmentation (twenty-seven [60%] of forty-five patients). The site of the elevated scapula was on the concavity of the scoliotic curvature in twenty-six (60%) of forty-three patients with scoliosis and on the convexity in seventeen. The combination of a congenitally elevated scapula and its occurrence on the convexity of an upper thoracic congenital scoliosis causes a substantial deformity because of an elevation of the shoulder line and impairment of shoulder function. These deformities usually require surgical treatment to correct the scoliosis and also to perform a distal displacement of the scapula in relation to the vertebral column. However, when the Sprengel deformity is on the concavity of the scoliosis, it often partially compensates for the cosmetic deformity caused by the elevation of the contralateral shoulder on the convexity of the scoliotic curve. This minimizes shoulder asymmetry and usually does not require reduction of the congenitally elevated scapula.

In conclusion, this study showed that congenital rib and chest wall anomalies occur most frequently on the concavity of a thoracic or thoracolumbar congenital scoliosis that is due to a unilateral failure of vertebral segmentation. These rib abnormalities, either simple or complex, do not appear to have an adverse effect on the size or the rate of progression of the curve. ■

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Lung Function in Congenital Kyphosis and Kyphoscoliosis

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Objective: To quantify the respiratory compromise in patients with a congenital kyphosis or kyphoscoliosis in whom the major deformity is the kyphosis.

Methods: Forty-one patients with congenital vertebral anomalies resulting in a kyphosis or kyphoscoliosis, in which the kyphosis was the major deformity, and requiring spine surgery were studied retrospectively. The preoperative respiratory function and radiographic spinal assessments were compared.

Results: Twenty-two patients (54%) had an impairment of respiratory function: 2 patients (5%) were severely affected, 8 patients (20%) were moderately affected, and 12 patients (29%) were mildly affected.

Conclusions: An increasing severity of kyphosis was associated with a significant increase in respiratory impairment ($P < 0.005$). A more cranial level of the kyphosis, especially above T10, had a significantly greater effect on respiratory impairment ($P < 0.001$). One untreated patient with a severe kyphosis (128 degrees) died from cor pulmonale.

Key Words: lung function, congenital kyphosis, congenital kyphoscoliosis

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A congenital kyphosis or kyphoscoliosis is an uncommon deformity in which there is an abnormal posterior convex angulation of a segment of the spine caused by developmental vertebral anomalies. These vertebral anomalies, which are present at birth, result in a localized impairment of anterior longitudinal spinal growth in the sagittal plane and an increasing deformity as the child grows to skeletal maturity.^{1,2} In some patients there is an asymmetrical impairment of anterior spinal growth resulting in a kyphoscoliosis but the major deformity in all of the patients in this study was the kyphosis.

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Previous studies have shown that a progressive infantile or congenital scoliosis which develops in early childhood and deforms the rib cage can result in an impairment of lung development and function.^{3–5} To the best of our knowledge, there has been no large series in the English language of the effect of a congenital kyphosis or kyphoscoliosis on lung function.

The purpose of this paper was to evaluate the effect of congenital kyphosis and kyphoscoliosis on respiratory function in relation to age as well as the site and severity of the spinal deformity in patients who were candidates for the surgery.

MATERIALS AND METHODS

We reviewed retrospectively the medical records, radiographs, and respiratory function tests performed on 41 patients with a congenital kyphosis or kyphoscoliosis before spinal surgery. These patients were part of a much larger study on the natural history and surgical management of congenital kyphosis and kyphoscoliosis.^{2,15} The normal sagittal profile of the spine was taken into consideration and all the patients had a sharp angular kyphosis and the associated scoliosis was a secondary component. None of these patients had significant congenital anomalies of the ribs or chest wall, lung hypoplasia, or congenital heart disease. The indication for surgical treatment was either the poor prognosis if left untreated or the size of their spinal deformity when first seen.¹⁵ Nine of the patients had pain and only one complained of breathlessness on exertion. Patients who had a kyphosis associated with a myelomeningocele, Scheuermann disease, neurofibromatosis, skeletal dysplasia, infection, or trauma were excluded.

There were 15 males and 26 females whose mean age at the time of their respiratory evaluation was 13 years 4 months (range 8 y 7 mo to 25 y). These measurements were performed as part of the assessment before spine surgery. It was not possible to perform accurate respiratory function tests on young children under the age of 8 years because of a failure to cooperate.

An additional patient who first presented at the age of 9 years with a 128-degree congenital thoracic kyphosis, died at the age of 28 years of respiratory failure and cor pulmonale. This patient is not included in the results because lung function tests were not available for review.

Radiographic Assessment

The type of vertebral anomaly causing the kyphosis was assessed on the antero-posterior and lateral spine

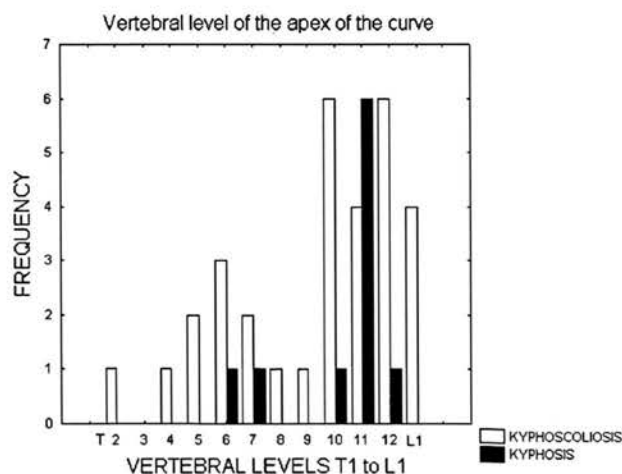
TABLE 1. Types of Vertebral Anomaly Producing Congenital Kyphosis or Kyphoscoliosis

Type	Subclassification	No. Patients
Failure of formation	Posterior hemivertebra or posterolateral quadrant vertebra	16
	Butterfly vertebra	5
	Wedge vertebra	3
Failure of segmentation	Anterior and anterolateral	6
Mixed		6
Unclassifiable		5

radiographs. The deformity was classified^{1,2} as type 1 (anterior failure of vertebral body formation) in 24 patients, type 2 (anterior failure of vertebral body segmentation) in 6 patients, and type 3 (mixture of failure of formation and segmentation) in 6 patients. Five patients could not be classified accurately because the vertebral anomalies were partially obscured by the severity of the deformity (Table 1).

The apex of the kyphosis was assessed on the lateral spine radiographs taken with the patient standing. Thirty-seven patients (90%) had apices of their curves in the thoracic region between T2 and T12 inclusive (Fig. 1). The mean extent of the measured kyphosis was over 5 vertebrae (range 3 to 9 vertebrae) and all patients had an angular deformity. An associated scoliosis was also present in the same region in 35 patients but the major deformity was the kyphosis.

The severity of the kyphosis was measured in all patients by the use of the modified Cobb method⁶ from the most sagittally tilted vertebra at either end of the deformity as seen on the lateral spine radiograph taken with the patient erect.

**FIGURE 1.** A Frequency histogram of the vertebral levels at which the apex of the curve is situated in kyphosis and kyphoscoliosis.

Pulmonary Function Tests

These tests were performed by respiratory technicians in cooperative children over the age of 8 years. Forced vital capacity (FVC) and forced expiratory volume in 1 second (FEV₁) were measured for each patient using a calibrated spirometer in an accredited pulmonary function laboratory. These tests were performed only once in the few months before surgery. The data were standardized in these patients to accommodate for shortening of the trunk by using arm span to predict the normal value for height.⁷ The percentage FVC and FEV₁ were calculated according to Polgar standards.

Statistical Analysis

The effect of *kyphoscoliosis* and its individual components, *scoliosis* and *kyphosis*, upon each of the 2 variables of respiratory function (FEV₁ and FVC) were tested using multiple linear regression. It was necessary first to show that the data were normally distributed, as this is a requirement of the regression test.

First, half normal plots of the data points with 95% confidence ellipses were constructed to identify outliers. Four such points were identified and removed from the series.

The residuals for the data points were then calculated and normal-probability graphs plotted using the "Statistica" program (Statsoft, USA). The distribution of the residuals was then tested against a computed distribution with the same mean and variance using the Shapiro-Wilk W test. The residuals for both of the dependent variables (FEV₁ and FVC) were found to fit a normal distribution. Hence, it was acceptable to proceed by using parametric statistical tests throughout.

In the test for the effect of *kyphoscoliosis* upon FEV₁ and FVC the measurements of the scoliotic Cobb angle and the kyphotic Cobb angle were considered together to assess their combined effect and also separately to discover their individual effects. A test was used, in which it was possible to combine 2 independent variables (SC and KY) with 2 dependent variables (FEV₁ and FVC). With combined independent variables the test lost sensitivity but reflected the biologic situation more accurately.

RESULTS

A spinal deformity was observed in all 41 patients before the age of 5 years but it was not until a mean age of 8 years 2 months (range 1 mo to 21 y) that spine radiographs were available for diagnosis; the mean kyphosis at this time was 63 degrees (range 12 to 135 degrees). Respiratory function tests were performed preoperatively at a mean age of 13 years 4 months (range 8 y 7 mo to 25 y) when the mean kyphosis had increased to 83 degrees (range 30 to 135 degrees). In all of these patients the kyphosis was sharp and angular. Thirty-five patients also had an associated scoliosis in the same region. In all of these patients the kyphosis was the major deformity and greater than the scoliosis whose mean size was 58 degrees (range 8 to 84 degrees).

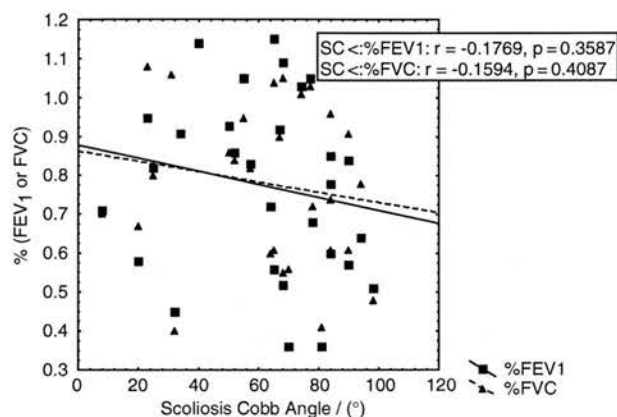


FIGURE 2. A regression plot of %FEV₁ and %FVC against the Cobb angle in patients with scoliosis.

According to the criteria of Johnston and Westgate,⁸ the respiratory function in 22 patients (54%) was abnormal. Two patients (5%) were classified as severely affected, 8 patients (20%) were moderately affected and 12 patients (29%) were mildly affected. The remaining 19 patients (46%) were within the range of normality.

Figure 1 is a histogram of the frequency at which the apices of the curves were distributed for kyphosis alone and for kyphoscoliosis. When the combined effect of scoliosis and kyphosis were considered as kyphoscoliosis: for FEV₁: $R = -0.43$ ($P = 0.090$); for FVC: $R = -0.41$ ($P = 0.110$); in other words when the effects of FEV₁ and FVC were considered together the combination of the 2 regressors had no significant effect on either FEV₁ or FVC.

Figures 2 and 3 show the results of the multiple regression tests using the angles of scoliosis and kyphosis separately as regressors. With scoliosis as the regressor for FEV₁ $R = -0.18$ ($P = 0.358$) and for FVC $R = -0.16$ ($P = 0.409$); in other words neither correlation was significant. With kyphosis alone as the regressor, for FEV₁ $R = -0.49$ ($P = 0.006$) and for the dependent

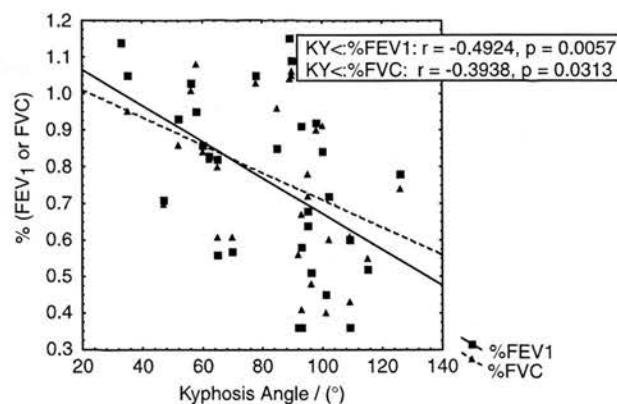


FIGURE 3. A regression plot of %FEV₁ and %FVC against the Cobb angle in patients with kyphosis.

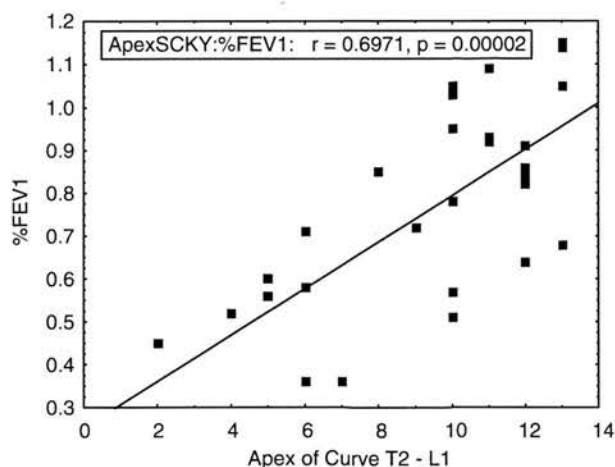


FIGURE 4. A regression plot of %FEV₁ against the position of the apex of the curve.

variable FVC $R = -0.39$ ($P = 0.031$); both of these were stronger correlations and both were significant.

There were strong correlations found when multiple regression was used to test the relationship between the apex of the kyphotic element of the curve and the results of the respiratory function tests in kyphoscoliosis. These relationships are shown in Figures 4 and 5.

The level of the apex of the kyphosis also had an effect on the severity of lung impairment. The higher the apex of the kyphotic deformity within the thoracic cage the worse the respiratory function and this was statistically significant (FVC, $R = 0.67$, $P < 0.001$; FEV₁, $R = 0.70$, $P < 0.001$). The mean predicted FVC for those patients whose kyphosis apex was above T10 (13 patients), compared with those whose apex was between T10 and T12 inclusive (24 patients) and those with their apex below T12 (4 patients) was 58%, 82%, and 97%, respectively. The mean predicted FEV₁ was 53%, 83%, and 100%, respectively.

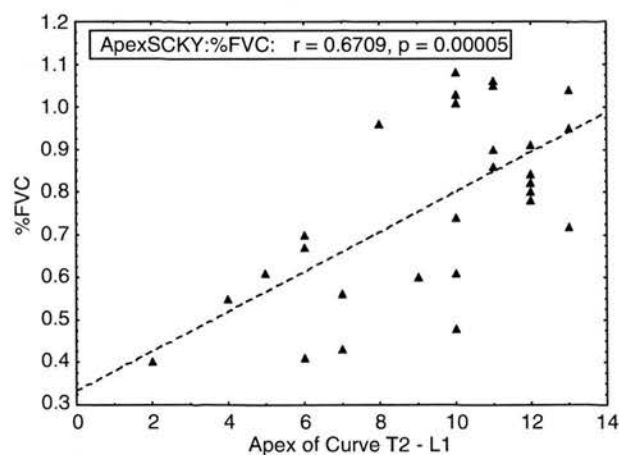


FIGURE 5. A regression plot of %FVC against the position of the apex of the curve.

Multiple logistic regression for respiratory impairment by level of curve apex and by severity of kyphosis showed apex level to be the most important variable for both percentage predicted FVC and FEV₁ ($P < 0.001$). Only percentage predicted FVC was significantly related to kyphosis severity ($P = 0.03$) when controlled by level of kyphotic apex. The percentage predicted FEV₁ was not significantly related ($P = 0.05$) to severity of kyphosis.

The ages at which respiratory function tests were performed ranged from 8 to 25 years. The size of the kyphosis at the time of testing varied greatly among these patients but there was no significant relationship between the age at testing and the severity of the kyphosis ($P = 0.06$) when analyzed by multiple logistic regression. Five patients aged between 8 and 10 years had a mean kyphosis of 80 degrees (range 30 to 121 degrees) and their mean predicted FVC was 56% of normal (range 31% to 84%). The 13 patients aged between 10 and 12 years had a mean kyphosis of 73 degrees (range 35 to 98 degrees) and their mean predicted FVC was 80% (range 47% to 116%). The 23 patients older than 12 years had a mean kyphosis of 91 degrees (range 47 to 135 degrees) and their mean predicted FVC was 78% (range 35% to 108%).

DISCUSSION

The results of this study indicate that lung function in young patients with a congenital kyphosis or kyphoscoliosis becomes more severely impaired as the angle of the kyphotic deformity increases ($P < 0.006$). A more cranial level of the apex of the kyphosis, especially above T10, results in a significantly greater respiratory impairment ($P < 0.001$).

This is a cross-sectional and not a longitudinal study and it is, therefore, not possible to say for an individual patient that the respiratory function becomes increasingly impaired as the kyphotic deformity increases. However, if we take into consideration all the patients, this study shows that with a more severe kyphosis, the respiratory function becomes more severely impaired. Multiple regression analysis showed that the correlation between the Cobb angle of the curve and the indices of respiratory function was due mainly to the kyphotic element of the deformity.

Congenital kyphosis, kyphoscoliosis, and scoliosis, are all part of a gradually blending spectrum of spinal deformities due to developmental vertebral anomalies. Patients whose major deformity is a kyphosis or a kyphoscoliosis are much less common than those with a pure scoliosis. With a congenital scoliosis occurring in the thoracic region there is a lateral curvature of the spine and an associated rib cage deformity. However, with an angular congenital thoracic kyphosis or kyphoscoliosis the spine is sharply angulated forwards compressing the ribs on both sides of the chest and also impairing movement of the diaphragm. This is likely to have a much greater effect on lung function than a pure scoliosis.

Early studies suggested that the number of lung alveoli continued to increase after birth until the age of 8 years.⁹ However, more recent studies have shown that normal alveolar multiplication is usually complete by the age of 2 years after which the alveoli continue to increase in size until thoracic growth is complete.¹⁰⁻¹² The development of the ancillary arteries and capillaries parallel the development of the alveoli. Reid and Davies studied 4 cadavers with severe rib cage deformity secondary to scoliosis occurring in early childhood. They showed that the alveoli failed to multiply and may even atrophy in the compressed developing lung.¹³

Progressive infantile or congenital scoliosis, which develops in early childhood and deforms the rib cage have been shown to result in an increasing impairment of lung function.³⁻⁵ It is, therefore, likely that a similar impairment will occur in patients with a congenital kyphosis or kyphoscoliosis developing at an early age. However, a spinal curvature with an associated rib cage deformity developing in adolescence, after the lungs are fully developed, will not have a significant effect on lung function unless there is an extreme deformity or an associated neuromuscular condition causing respiratory muscle paralysis.

Owange-Iraka et al³ studied the respiratory function in 24 patients with congenital scoliosis and 30 patients with infantile idiopathic scoliosis. They found that the lung vital capacity (VC) in both groups of patients deteriorated progressively as the scoliosis progressed. However, for any given Cobb angle of the scoliosis, the loss in VC was approximately 15% greater in congenital scoliosis than in infantile idiopathic scoliosis. The 24 patients with congenital scoliosis had their lung function assessed at a mean age of 11 years (range 4 to 26 y) when the mean Cobb angle of the scoliosis was 65 degrees (range 16 to 99 degrees) and the mean VC was found to be 47% of normal (range 13% to 68%). Muirhead and Conner⁴ in a study of 25 patients with congenital scoliosis also found a significant correlation between diminished VC and severity of curvature. They assessed their patients at a mean age of 12 years when the mean Cobb angle was 71 degrees (range 43 to 130 degrees) and the mean FVC was 67%. In contrast, Day et al⁵ found normal lung function in 11 untreated patients with congenital scoliosis whose mean age was 11.6 years but their mean Cobb angle of curvature was much smaller at 34 degrees (range 16 to 58 degrees).

In our study, all 41 patients with a congenital kyphosis or kyphoscoliosis were noted to have a spinal deformity before the age of 5 years. However, spine radiographs were not available for diagnosis until a mean age of 8 years 2 months (range 1 mo to 21 y) when the mean kyphosis was 63 degrees (range 12 to 135 degrees). The apex of the kyphosis in 37 of these patients (90%) was in the thoracic region between T2 and T12 inclusive. All of these patients had a progressive angular kyphosis with an associated rib cage deformity. It is likely that this severity of deformity developing in the thoracic region before the age of 8 years would have had a detrimental

effect on the normal development of the lung. However, a limitation of this study is that it was not possible to perform accurate standard lung function tests on children younger than 8 years because of a failure to comply. Respiratory function tests were not performed on our patients until a mean age 13 years 4 months (range 8 y 7 mo to 25 y) when the mean kyphosis was 83 degrees (range 35 to 135 degrees) immediately before spine surgery. Ideally, it would have been much better to have operated on these children before the age of 5 years when they had smaller curves. However, many of the patients in this study were referred late to our tertiary spinal deformity center when they were much older with larger spinal deformities.

There is a large error of estimation inherent in predicting VC and Johnson and Westgate⁸ arbitrarily classified patients with up to 20% reduction in VC as normal; those with values between 60% and 80% have mild restriction; those between 40% and 60% have moderate restriction; and those below 40% have severe restriction. According to these criteria the effect on lung function in our patients was severe in 2 patients (5%), moderate in 8 patients (20%), mild in 12 patients (29%), and nil in 19 patients (46%).

To prevent the development of a severe spinal deformity it has been shown that a congenital kyphosis or kyphoscoliosis is best treated prophylactically before the age of 5 years by a posterior spinal fusion to balance the growth of the spine before the kyphosis exceeds 40 degrees.^{14,15} This relatively simple procedure produces a posterior spinal tether, which either stabilizes the deformity or in the presence of continuing anterior longitudinal growth provides the opportunity for gradual correction.

Unfortunately, it is not possible to perform accurate respiratory function tests on young children under the age of 5 years due to a failure to cooperate. However, it is unlikely that there would be a significant respiratory impairment in these young children with small curves and the indication for surgery is to prevent increasing spinal deformity and distortion of the rib cage. It is well established that it is the distortion of the rib cage, which causes the impairment of lung growth and development, and if the rib cage is not deformed, one could conclude that the respiratory function would be maintained.

In our study the earliest lung function tests were performed on 5 patients between the ages of 8 and 10 years when their mean kyphosis was 91 degrees (range 60 to 102 degrees) and their mean predicted FVC was 46% of normal (range 43% to 84%). All had a significant deformity of the rib cage. Obviously, these patients would have been much better treated prophylactically by spinal surgery at a much earlier age.

A kyphosis which does not reduce to less than 50° especially in an older patient will require a much more complex 2-stage procedure to perform an anterior spinal release and arthrodesis with strut grafting, followed by posterior arthrodesis with instrumentation (if possible) to correct and stabilize the deformity.^{15,16}

In these older children with larger deformities it is much more likely that there would be an impairment of respiratory function. However, the main indication for surgery is to prevent an increasing kyphosis leading to spinal cord compression and paraplegia.

In our study 36 patients (88%) had a kyphosis over 50 degrees (range 52 to 135 degrees) and would have benefited from anterior and posterior spine surgery. Their mean predicted FVC was 75% (range 31% to 116%) and the mean FEV₁ was 74% (range 26% to 116%). Anterior spinal surgery in these patients requires a thoracotomy or thoracoabdominal approach to the spine by dividing the diaphragm. This would have a significant adverse effect on lung function during the postoperative period in patients whose lung function had already been compromised by their chest deformity.

A limitation of this study is that lung function tests were not performed postoperatively. However, there was a wide spectrum of operations performed on these patients; some had prophylactic posterior procedures at an early age, others with more severe deformities had a posterior spinal fusion at a later age and some had combined anterior and posterior surgery. This introduces many operative variables with regard to postoperative lung function and the number of patients in each subgroup would be small.

In conclusion, it is our opinion, that surgical treatment for a progressive congenital kyphosis or kyphoscoliosis is indicated at an early age not only to prevent severe spinal deformity and possible neurologic complications^{1,2} but also to prevent the adverse effect on lung development and function caused by an increasing constriction of the rib cage and impairment of diaphragmatic movement.

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Goldenhar-Associated Conditions (hemifacial microsomia) and Congenital Deformities of the Spine

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Study Design. We performed a retrospective study of 668 consecutive patients with congenital spine deformities.

Objectives. To determine the prevalence of Goldenhar conditions in patients with congenital spine deformities, and describe the types of vertebral abnormalities and their treatment.

Summary of Background Data. The craniofacial malformations described by Goldenhar can be associated with congenital vertebral anomalies. This nonrandom association represents unilateral errors in the morphogenesis of the spine, the first and second branchial arches.

Methods. The medical records were reviewed, and patients with Goldenhar conditions were identified. The vertebral anomalies causing spine deformity were detected on radiographs. The type and site of the craniofacial abnormalities, other musculoskeletal deformities, and systemic anomalies were recorded.

Results. There were 14 patients, including 7 males and 7 females, who had Goldenhar conditions. Thoracic scoliosis was the most common deformity, occurring in 10 patients (71.5%). Of these patients, 8 had an isolated hemivertebra, and the remaining 2 had a unilateral unsegmented bar with contralateral hemivertebra at the same level. There was only 1 patient with lumbar scoliosis caused by a hemivertebra. The side of vertebral anomaly correlated with that of hemifacial microsomia in 5 of 11 patients with scoliosis or kyphoscoliosis. Thoracolumbar kyphosis occurred in 4 patients, including 2 who had posterior hemivertebrae, 1 who had wedge vertebrae, and the remaining who had an anterior unsegmented bar. Thoracolumbar kyphoscoliosis occurred in only 1 patient because of a posterolateral quadrant vertebra. Klippel-Feil syndrome occurred in 6 patients (42.8%). There were 8 patients (57%) who underwent surgical treatment at a mean age of 9.8 years (range 2.9–19). Four patients had undergone a combined anterior-posterior, and 4 had a posterior-only spine arthrodesis.

Conclusions. The prevalence of Goldenhar-associated conditions in patients with congenital spine deformities was 2%. Failures of vertebral segmentation were the most frequent abnormality in the cervical spine, whereas failures of vertebral formation most commonly occurred in the thoracic or thoracolumbar spine.

Key words: congenital scoliosis, kyphosis, kyphoscoliosis, Goldenhar syndrome, hemifacial microsomia, oculoauriculovertebral dysplasia, facio-auriculo-vertebral spectrum.
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Congenital deformities of the spine are a gradually blending spectrum of deformities, ranging from a scoliosis through kyphoscoliosis to a pure kyphosis. These deformities are caused by an asymmetrical failure of development of ≥ 1 vertebra as a consequence of a unilateral failure of formation or segmentation of the primitive somites. The presence of a congenital vertebral anomaly can result in a localized imbalance in the longitudinal growth of the spine and an increasing spinal curvature, which continues to progress until skeletal maturity.^{1–4} On rare occasions, these vertebral abnormalities may be associated with a unilateral failure of formation of the face and ear; this combination of anomalies is best described as hemifacial microsomia.

Hemifacial microsomia is a common birth defect caused by a unilateral arrest or dysplastic development of the first and second branchial arches, and the intervening first pharyngeal pouch and branchial cleft. This results in unilateral hypoplastic development of the external and middle ear, ramus and condyles of the mandible, zygoma, and facial muscles.^{4–6} The phenotype of this condition is highly variable, and many transitional forms exist.

In 1952, Goldenhar⁷ reported 3 patients with what he considered to be a new syndrome consisting of a combination of unilateral auricular appendages, auricular fistulas, and ocular epibulbar dermoids combined with a unilateral underdevelopment of the craniofacial bony structures. Goldenhar did not describe an association with vertebral abnormalities or spine deformity. Since Goldenhar's original description, other associated anomalies have been added, including upper eyelid colobomas, subconjunctival lipomas, hearing loss, micrognathia, cleft or high-arched palate, and congenital heart disease.⁸ However, it was not until 1963, when Gorlin *et al*⁹ recognized that these Goldenhar-related conditions could also be associated with developmental anomalies of the vertebral column. They suggested the designation oculoauriculovertebral dysplasia to include the 3 predominant groups of malformations.

Smith¹⁰ used the term facio-auriculo-vertebral spectrum to include both hemifacial microsomia and Goldenhar syndrome, along with the phenotypic spectrum of similar conditions. The existence of a multitude of transitional forms between hemifacial microsomia and Gold-

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enhar syndrome have suggested that no valid distinction could be made between these 2 conditions, which may simply represent gradations in severity of a similar error in morphogenesis (Table 1).^{5,25-27}

There have been extensive reports regarding the facial and auricular anomalies in Goldenhar-related conditions,⁷⁻⁹ but very little information is available in the orthopedic literature regarding the associated spine problems or their frequency. The purpose of our study was 2-fold. First, we wanted to determine the prevalence of hemifacial microsomia in patients with congenital spinal deformities and describe the types of vertebral anomalies that can occur in patients with this condition, as well as the necessity for treatment. Second, we wanted to increase awareness of a spectrum of conditions in which the craniofacial anomalies may be obvious and easily appreciated at birth in contrast to the vertebral malformations, which can be frequently overlooked until a clinically recognizable spinal deformity develops in later life.

Materials and Methods

We reviewed the medical records and spine radiographs of 668 consecutive patients with a congenital spine deformity seen at

our institution between 1960 and 2003. A total of 532 patients had a scoliosis (80%), 95 had a kyphoscoliosis (14%), and 41 had a kyphosis (6%). Patients who had myelomeningocele, Scheuermann disease, neurofibromatosis, skeletal dysplasia, infection, or trauma were excluded.

We identified 14 patients, including 7 males and 7 females, with a diagnosis of hemifacial microsomia. All patients had more than 2 years of follow-up for their spine deformity. The type and site of craniofacial abnormalities were recorded on the clinical examination. The presence of associated systemic anomalies and musculoskeletal deformities was also documented. The vertebral anomalies causing the spine deformity were detected on anteroposterior and lateral spine radiographs. The vertebral anomalies and type of spine deformity were categorized according to the classification proposed by Winter^{1,2} and McMaster^{3,4} *et al* into failures of vertebral segmentation, failures of vertebral formation, and mixed anomalies producing a congenital scoliosis, kyphoscoliosis, or kyphosis.

The site of the curvature was defined according to the classification proposed by the Scoliosis Research Society²⁸: cervicothoracic (apex C7 or T1), thoracic (apex T2-T11), thoracolumbar (apex T12 or L1), lumbar (apex L2-L4), and lumbosacral (apex L5 or below). The spinal radiographs were examined for any evidence of intraspinal anomaly or a Klippel-Feil syndrome. Magnetic resonance imaging of the

Table 1. Goldenhar-Related Conditions (hemifacial microsomia)

Incidence	HM 1:5-6000 ¹¹	OAVD 1:3500-5000 Live Births ¹⁰	OAVD Minimum 1:45000 ¹²			
Inheritance	Most commonly sporadic cases ⁹	Occasionally autosomal dominant and autosomal recessive trait ¹³⁻¹⁸				
Pathogenesis	Vascular theory (a fetal hemorrhage occurring into the differentiating branchial arch tissues leads to the formation of a hematoma that can disrupt the vascularity of part of the face and result to an ischemic necrosis) ¹⁹	Genetic causes linked to the vascular disruption ²⁰	Primary mesodermal deficiency ²¹	Disturbed neural crest development common in both OAVD and CHARGE association ²²	Deficiency in neural crest cell migration, in mesodermal formation or defective interaction between neural crest cells and mesoderm may cause defects of blastogenesis ²³	Poorly controlled maternal diabetes interfering with neural crest cell migration ²⁴
Clinical Features	Facial: unilateral hypoplasia, micrognathia, cleft or high-arched palate	Auricular: unilateral deformity of the external ear, unilateral appendages, fistulas, hearing loss, cartilaginous preauricular tags, atresia of the external auditory canal	Ocular: epibulbar dermoids, upper eyelid colobomas, subconjunctival lipomas, antimongoloid slant	Spine: hemivertebra, unsegmented bar, cuneiform vertebra, Klippel-Feil syndrome, platybasia, occipitalization of the atlas, spina bifida, lumbarization of the sacrum, aplasia of the sacral vertebrae	Other musculoskeletal: DDH, radial limb anomalies, triphalangeal thumbs, clinodactyly, clubfoot, calcaneovalgus, syndactyly of the toes, short stature, rib anomalies	Systemic: congenital heart disease, esophageal atresia, tracheoesophageal fistula, genitourinary abnormalities, lung hypoplasia

DDH indicates developmental dysplasia of the hip; HM, hemifacial microsomia; OAVD: oculoauriculovertebral dysplasia.

whole spine or myelography was performed in 4 patients as part of the preoperative assessment to evaluate the intraspinal content and exclude the presence of spinal dysraphism.

■ Results

The clinical findings in our patient population, including craniofacial and systemic manifestations of the condition, as well as information on the type of spine deformity and the treatment are shown in Tables 2 and 3. All 14 patients were referred to our spine clinic for evaluation of their spinal deformity. Mean age at presentation was 8 years (range 0.1–19).

None of the patients included in our study had a family history of Goldenhar-associated conditions. There was no history of maternal diabetes during pregnancy, and there were no affected siblings. There

Table 2. Description of Clinical Findings in Our Patient Population

Patient No. (sex)	Facial	Auricular	Ocular	Skeletal	Systemic
1 (M)	(Lt) hemifacial hypoplasia-hypoplastic (Lt) maxilla and mandible	(Lt) ear deformed-absent external auditory meatus-anterior accessory appendage-(Lt) deafness	Antimongoloid slant	Short stature	Esophageal atresia-tracheoesophageal fistula
2 (F)	(Lt) hemifacial hypoplasia-hypoplastic (Lt) maxilla and mandible	(Lt) ear deformed-(Lt) deafness	Abnormal (Lt) tear duct	Klippel-Feil syndrome (C ₄ -C ₆ fusion)	Dextrocardia
3 (M)	(Lt) hemifacial hypoplasia-hypoplastic (Lt) maxilla and mandible	(Lt) ear deformed-absent external auditory meatus-(Lt) deafness		Short stature-(Lt) radial clubhand-(Lt) clubfoot	Dextrocardia-undescended testicle
4 (M)	(Rt) hemifacial hypoplasia-hypoplastic (Rt) maxilla and mandible	(Rt) ear deformed-severe (Rt) hearing impairment	Antimongoloid slant	Short stature	
5 (F)	(Rt) hemifacial hypoplasia-hypoplastic (Rt) maxilla and mandible	(Rt) ear deformed-severe (Rt) hearing impairment	Ptosis of the (Rt) eye		
6 (F)	(Lt) hemifacial hypoplasia-hypoplastic (Lt) maxilla and mandible	(Lt) ear deformed-absent external auditory meatus-failure of formation of the pinna-severe (Lt) hearing impairment			
7 (F)	(Lt) hemifacial hypoplasia-hypoplastic (Lt) maxilla and mandible	(Lt) ear deformed-severe (Lt) hearing impairment	Antimongoloid slant	Short stature-(Lt) DDH-(Rt) radial clubhand	Atrial septal defect
8 (M)	(Rt) hemifacial hypoplasia-hypoplastic (Rt) maxilla and mandible	(Rt) ear deformed-severe (Rt) hearing impairment		Bilateral per cavus-Klippel-Feil syndrome (C ₂ -C ₃ fusion)	
9 (M)	(Rt) hemifacial hypoplasia-hypoplastic (Rt) maxilla and mandible	(Rt) ear deformed-absent external auditory meatus-(Rt) deafness		Short stature-(Rt) per cavus-LLD (Rt leg short)-Klippel-Feil syndrome (C ₆ -C ₇ fusion)	
10 (M)	(Rt) hemifacial hypoplasia-hypoplastic (Rt) maxilla and mandible	(Rt) ear deformed-(Rt) deafness		Klippel-Feil syndrome (C ₂ -C ₃ fusion-hypoplastic (Rt) thumb)	Hypospadias, hypoplastic (Rt) lung, dextrocardia, absent (Rt) kidney, (Lt) megaureter
11 (F)	(Lt) hemifacial hypoplasia-hypoplastic (Lt) maxilla and mandible	(Lt) ear deformed-(Lt) deafness		Klippel-Feil syndrome (C ₂ -C ₃ fusion)-multiple rib fusions on the concavity of scoliosis	
12 (F)	(Rt) hemifacial hypoplasia-hypoplastic (Rt) maxilla and mandible-(Rt) facial palsy	(Rt) ear absent-absent external auditory meatus-(Rt) deafness		Generalized muscle hypotonia-joint hyperlaxity	
13 (M)	(Lt) hemifacial hypoplasia-hypoplastic (Lt) maxilla and mandible	(Lt) preauricular skin tag-normal hearing on the (Lt), but low frequency loss on the (Rt)	(Lt) epibulbar dermoid cyst-(Lt) blindness-antimongoloid slant		
14 (F)	(Rt) hemifacial hypoplasia-hypoplastic (Rt) maxilla and mandible-nasal dermoid cyst-cleft palate	(Rt) ear deformed-severe (Rt) hearing impairment	(Bt) brainstem nystagmus	Klippel-Feil syndrome (C ₂ -C ₄ fusion)-(Rt) DDH-(Rt) accessory thumb	

Bt indicates bilateral; C, cervical; DDH, developmental dysplasia of the hip; F, female; LLD, leg length discrepancy; Lt, left; M, male; Rt, right.

Table 3. Description of Spinal Deformities in Our Patient Population

Patient No. (sex)	Type of Vertebral Anomaly	Type of Deformity	Age at Presentation (ys)	Neurology	Imaging Studies	Treatment	Follow-Up (ys)
1 (M)	T ₁₁ posterolateral quadrant vertebra	(Rt) TL kyphoscoliosis	5.8	No	Normal MRI whole spine	Anterior spine fusion (vascularized rib graft)-posterior instrumented spine fusion	18
2 (F)	T ₄ , L ₃ hemivertebrae	(Rt) T scoliosis, (Rt) L scoliosis	1	No	N/A	Observation	15
3 (M)	T ₃ , T ₅ , T ₉ , T ₁₁ , L ₃ hemivertebrae, T13 butterfly vertebra	(Rt) T scoliosis	0.4	No	N/A	Posterior spine fusion-no instrumentation	17
4 (M)	T ₅ , T ₁₀ hemivertebrae	(Rt) mid-T scoliosis, (Rt) lower T scoliosis	15.6	No	N/A	Observation	2
5 (F)	T ₃ -T ₄ block vertebra, T ₅ hemivertebra	(Rt) T scoliosis	1.9	No	N/A	Posterior spine fusion-no instrumentation	14
6 (F)	T ₇ hemivertebra	(Lt) T scoliosis, (Rt) TL-SC scoliosis	15	No	N/A	Observation	N/A (seen once at maturity)
7 (F)	T ₆ hemivertebra	(Rt) T scoliosis	0.1	No	N/A	Observation	20
8 (M)	Unilateral unsegmented bar T ₃ -T ₆ , T ₄ , T ₆ hemivertebrae, T ₈ butterfly vertebra	(Lt) T scoliosis, (Rt) TL-SC scoliosis	16.9	No	N/A	Observation	5
9 (M)	T ₁₂ posterior hemivertebra	TL kyphosis	19	No	Normal myelogram	Anterior spine fusion (fibular/rib strut grafts)-posterior instrumented spine fusion	2
10 (M)	T ₁ -T ₂ block vertebra, anterior unsegmented bar T ₁₃ -L ₁	(Lt) upper T scoliosis, TL kyphosis	12.8	No	N/A	Observation	8
11 (F)	T ₂ hemivertebra	(Rt) T scoliosis	1	No	N/A	Posterior spine fusion-no instrumentation	10
12 (F)	T ₁₀ anterior wedge butterfly vertebra, T ₁₁ anterior wedge vertebra	TL kyphosis	10	No	Normal MRI whole spine	Anterior spine fusion(vascularized rib graft)-posterior spine fusion-no instrumentation	5
13 (M)	T ₁₂ posterior hemivertebra	TL kyphosis	1.9	No	N/A	Posterior spine fusion-no instrumentation	12
14 (F)	Unilateral unsegmented bar T ₁ -T ₃ , T ₂ hemivertebra	(Lt) T scoliosis, (Rt) T-SC scoliosis	10.7	No	Normal MRI whole spine	Anterior spine fusion-posterior instrumented spine fusion	2

F indicates female; L, lumbar; Lt, left; M, male; N/A, not available; Rt, right; SC, structural compensatory; T, thoracic; TL, thoracolumbar.

was an equal distribution of patients according to the side of hemifacial hypoplasia. All patients had auricular deformities on the side of the facial hypoplasia (Figures 1A, B).

Spine Deformity

There were 10 patients (71.5%) who had a congenital thoracic scoliosis, and 2 of these had a second congenital thoracic or lumbar scoliotic curvature (Figure 2). Three patients had a structural compensatory thoracic or thoracolumbar curve below the primary congenital scoliosis. There was 1 patient who had a combination of a small congenital scoliosis caused by a block vertebra in the thoracic region and a congenital kyphosis caused by an anterior unsegmented bar in the thoracolumbar region. Three

patients had a pure congenital thoracolumbar kyphosis caused by a posterior hemivertebra (2 patients) (Figure 3) or anterior wedge vertebrae (1 patient). There was 1 patient who had a thoracolumbar kyphoscoliosis caused by a posterolateral quadrant vertebra.

Treatment

Eight patients (57%) underwent surgical treatment at a mean age of 9.8 years (range 2.9–19). The aim of the surgery was to stabilize primarily and correct partly the deformity. Of these patients, 4 had undergone a combined anterior-posterior spine arthrodesis, including 2 with kyphosis, 1 with kyphoscoliosis, and 1 with scoliosis. A vascularized rib autograft was used with or without additional fibular struts in the 3 patients

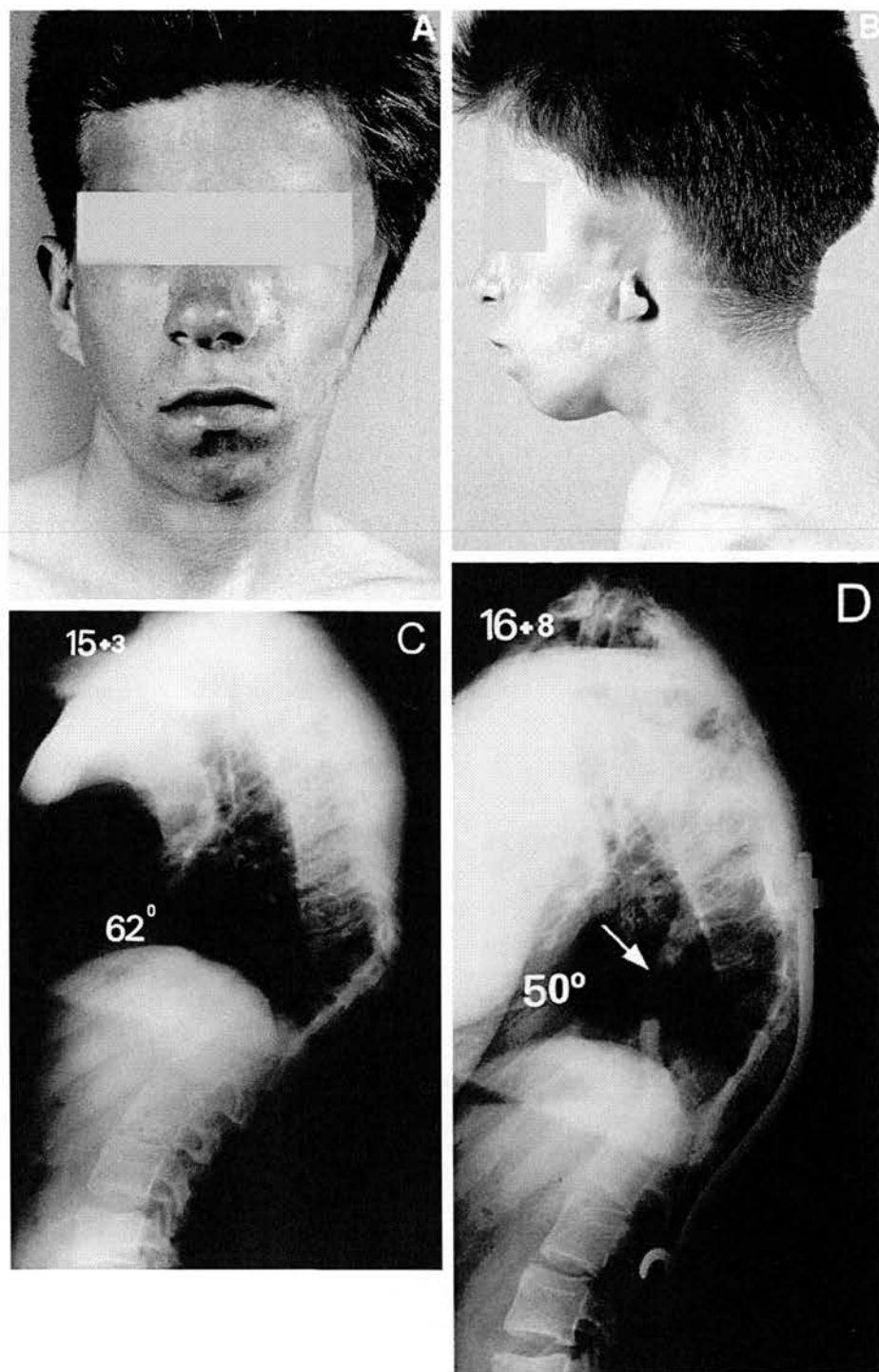


Figure 1. Anteroposterior (A) and lateral (B) clinical photographs in patient No. 1 show left hemifacial hypoplasia and a markedly deformed ipsilateral ear with absent external auditory meatus and an anterior accessory appendage. Lateral spine radiographs (C and D) in the same patient indicate improvement of the thoracolumbar kyphosis following a combined anterior-posterior spinal arthrodesis with the use of a vascularized rib autograft (white arrow) and posterior instrumentation.

with congenital kyphosis or kyphoscoliosis to provide anterior structural support (Figures 1C, D). The remaining 4 patients had undergone only a posterior spine arthrodesis, including 3 with scoliosis and 1 with kyphosis (Figure 3).

The patients were all mobilized in a spinal jacket until good radiographic evidence of a bony fusion was obtained. There was no evidence of pseudarthrosis or loss of correction at the latest follow-up and no neurologic deficits. Ten patients were observed into skeletal matu-

riety. Three patients who have been surgically treated are still under observation. The mean follow-up for this group of patients was 10 years (range 2–20). There was 1 patient with a congenital scoliosis who was seen only once at maturity and required no surgical treatment (patient No. 6).

■ Discussion

Developmental deformities of the vertebrae can result in an imbalance in the longitudinal growth of the spine and an

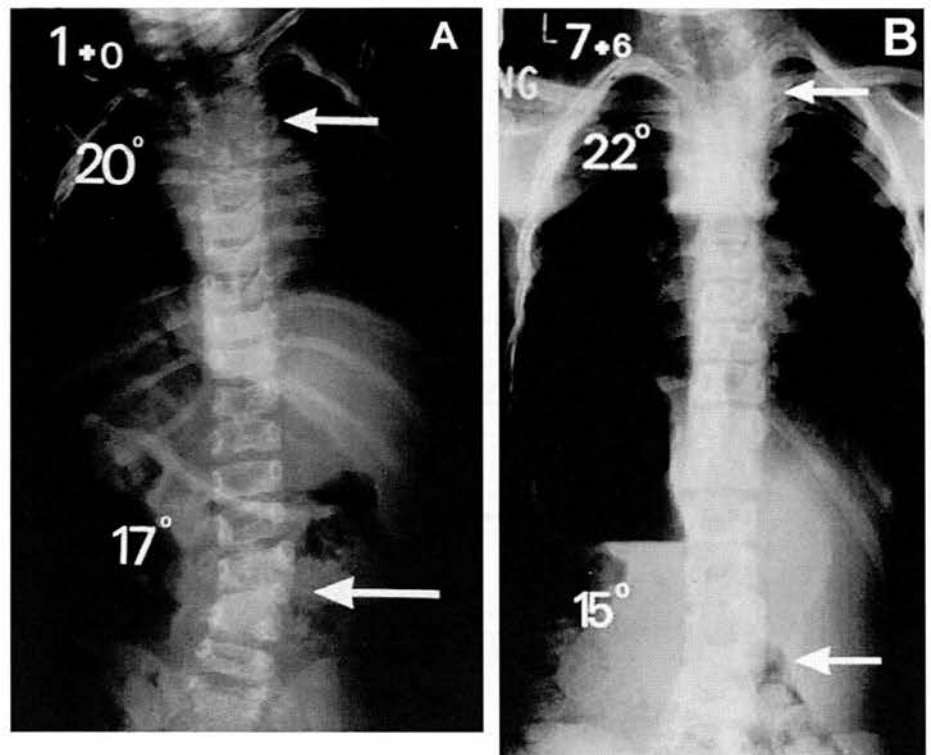


Figure 2. Anteroposterior spine radiographs in patient No. 2 show 2 ipsilateral hemivertebrae (white arrows) producing a thoracic and a lumbar scoliosis (A) with no evidence of progression over 6.5 years (B). Note the presence of dextrocardia.

increasing deformity as the child grows to maturity.¹⁻⁴ In 1963, Gorlin *et al*⁹ were the first to recognize that the congenital facial, ophthalmic, and auricular malformations, earlier described by Goldenhar,⁷ could also be associated with congenital anomalies of the vertebrae. The significant stages in the embryologic development of both the spine and skull occur during the first 6 weeks of intrauterine life. Therefore, craniofacial and vertebral anomalies are likely to be the result of a common pathogenetic mechanism occurring at the same stages of embryologic development. This nonrandom association of anomalies represents unilateral errors in the morphogenesis of the spine, as well as in the first and second branchial arches. However, it is likely that this association has been underestimated because not all developmental vertebral abnormalities result in a clinically obvious spinal deformity and, therefore, remain unrecognized.

We reviewed 668 consecutive patients with congenital deformities of the spine primarily referred to a spinal clinic because of a spine deformity and found that the prevalence of hemifacial microsomia was 2% (14 patients). A thoracic scoliosis was the most common deformity, occurring in 10 patients (71.5%). Of these patients, 8 had an isolated hemivertebra, and the remaining 2 had a unilateral unsegmented bar with contralateral hemivertebra at the same level. There was only 1 patient with a lumbar scoliosis, and this was caused by a hemivertebra. The side of the vertebral anomaly correlated with that of the hemifacial hypoplasia in 5 of the 11 patients who had a scoliosis or kyphoscoliosis. A thoracolumbar kyphosis occurred in 4 patients; 2 had a posterior hemivertebra, 1 had wedge vertebrae, and the remaining patient had an anterior

unsegmented bar. A thoracolumbar kyphoscoliosis occurred in only 1 patient and was caused by a posterolateral quadrant vertebra.

In contrast, Gibson *et al*⁵ reviewed 35 patients who were primarily diagnosed as having a Goldenhar syndrome and found that 60% had vertebral anomalies. Defects of vertebral segmentation occurred most frequently in the cervical spine (7 patients). Conversely, defects of formation were the most common pattern of congenital vertebral abnormality in the thoracic spine and occurred in 17 patients. Of these patients, 9 had a thoracic scoliosis caused by a hemivertebra, 2 had a thoracolumbar kyphosis caused by an anterior unsegmented bar, and 1 had a butterfly vertebra and a semi-segmented hemivertebra producing an upper thoracic kyphoscoliosis. Surgical correction of the deformity was required in 3 patients, including 2 patients with a congenital thoracic scoliosis secondary to multiple hemivertebrae and 1 with a thoracic kyphoscoliosis.

Klippel-Feil syndrome occurred in 6 of our patients (42.8%). This is higher than the reported incidence of 25% in patients with a congenital scoliosis alone.^{29,30} None of our patients had a failure of vertebral formation in the cervical spine (*i.e.*, hemivertebra). Congenital rib fusions occurred on the concavity of a thoracic scoliosis in 1 patient. Manifestations of Goldenhar-related conditions reported in previous studies are: occipitalization of the atlas³¹; cuneiform vertebra; platybasia (with an incidence of 12%); partial or complete cervical synostosis of ≥ 2 vertebrae; hemivertebrae^{5,8,32-35}; spina bifida^{8,11,32,33}; lumbarization of the sacrum or aplasia of the sacral vertebrae; and rib anomalies.⁵

Musculoskeletal abnormalities affecting areas other than the spine occurred in 4 of our patients (28.6%): devel-

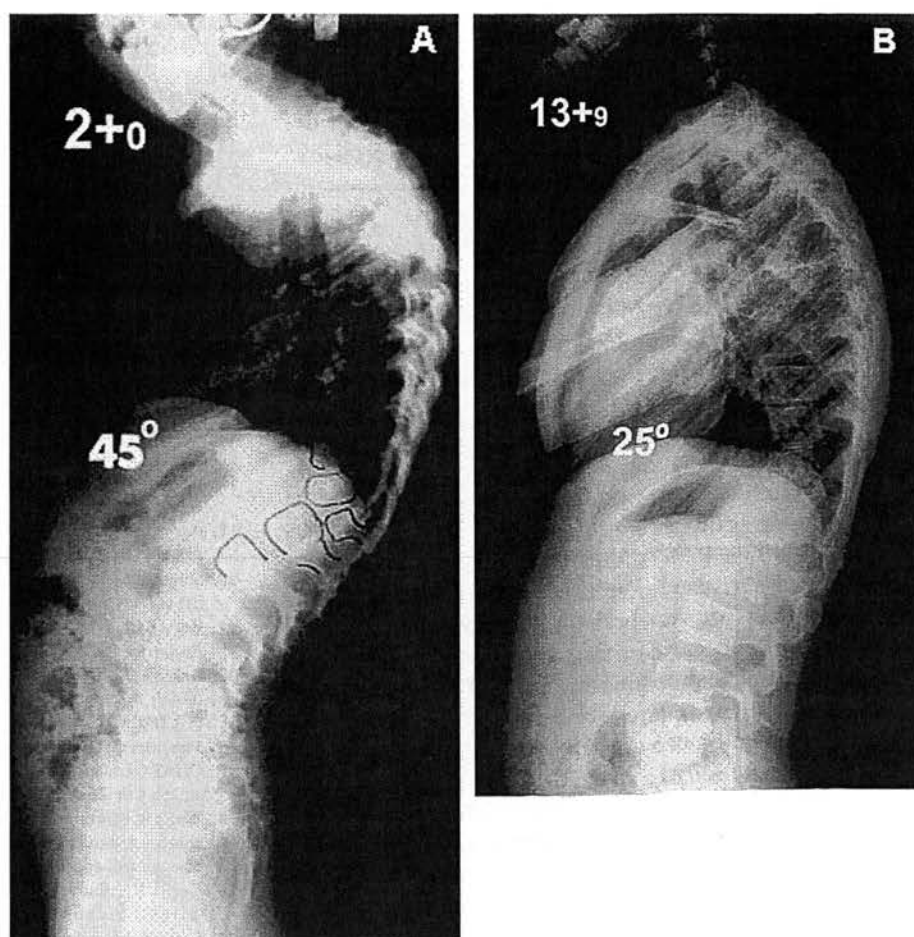


Figure 3. Lateral radiograph of the spine in patient No. 13 indicating a posterior hemivertebra at the T12 level producing a thoracolumbar kyphosis (A). The patient underwent a localized posterior spinal arthrodesis with iliac crest autologous bone, which produced a tethering effect and allowed for gradual correction of the kyphosis with further anterior vertebral growth (B).

opmental dysplasia of the hip in 2 patients, radial clubhand in 2, clubfoot in 1, and 1 patient had a unilateral accessory thumb. There were 5 patients (35.7%) who had short stature, which exceeded that attributable to the malformed vertebral segments alone. Other studies have shown isolated occurrences of clubfoot,^{11,32} congenital dislocation of the hip,³⁵ radial limb anomalies,^{34,36} unilateral radial hypoplasia with absence of the thumb, triphalangeal thumbs, clinodactyly, calcaneovalgus, syndactyly of the toes, and short stature.⁵

The craniofacial features of hemifacial microsomia include unilateral deformity of the external ear, hypoplasia of the ipsilateral half of the face, and epibulbar dermoids. Coloboma of the upper eyelid can also occur. The ear anomalies range from cartilaginous preauricular tags, atresia of the external auditory canal, and abnormalities in the shape and size of the external auricle to anotia. In our study, all patients had unilateral facial hypoplasia and ipsilateral malformation of the ear. However, there was only 1 patient with an epibulbar dermoid cyst, and none had a coloboma of the eyelid.

Congenital heart disease occurred in 4 of our patients (28.6%), which is higher than the reported incidence of 10% in patients with congenital scoliosis alone.³⁷ Dextrocardia was present in 3 of our patients (21.4%), which is also higher than the reported prevalence

of 1.5% in a retrospective review of 5539 fetal echocardiograms and 0.7% among 1039 patients with congenital heart diseases.^{38,39} None of our patients had neurologic abnormalities. Imaging studies of the spinal cord and cauda equina were performed in 4 patients and showed normal development.

Esophageal atresia and a tracheoesophageal fistula were present in 1 of our patients. Sutphen *et al*⁴⁰ reported 60 patients with the oculoauriculovertebral spectrum and found 3 who had tracheoesophageal fistula with or without esophageal atresia. Abnormalities of the genitourinary system occurred in only 1 of our patients, which is less than the reported incidence of up to 25% in patients with congenital scoliosis alone.^{41,42}

Spinal surgery is frequently necessary for patients with congenital deformities of the spine. This surgery is best performed at an early stage with the object of balancing the growth of the spine and preventing progressive deformity according to the indications described by Winter^{1,2} and McMaster^{3,4} *et al*. Of the 14 patients in our study, 8 (57%) required a spine arthrodesis. There were 4 patients seen before the age of 2 years, and they required prophylactic posterior surgery alone (Figure 3). However, the remaining 4 patients were seen at a latter stage when it was necessary to perform more complex combined anterior and posterior salvage procedures

(Figures 1C, D), which are associated with an increased risk of medical and neurologic complications. If the congenital vertebral anomalies in these patients had been recognized at a younger age with a lesser degree of deformity, more simple surgery could have been effective in controlling their deformity.

■ Conclusions

Goldenhar-associated conditions cover a nonexclusive phenotypic spectrum of pathologic entities, which are related to defects of morphogenesis occurring at the same stages of embryologic development. There is a well-recognized association between the craniofacial and musculoskeletal manifestations of these conditions. Congenital anomalies of the vertebral column producing a spine deformity comprise the most common orthopedic problem in these patients. This study has shown that failures of vertebral segmentation were the most frequent abnormality in the cervical spine, whereas failures of vertebral formation most commonly occurred in the thoracic or thoracolumbar spine. Patients with hemifacial microsomia are diagnosed at birth and should be routinely referred for evaluation by a spine specialist. Identification of a congenital vertebral anomaly will allow for recognition of a spinal deformity at an early stage when appropriate surgery can produce better results, reduce the risk of potential complications, and prevent unnecessary morbidity.

■ Key Points

- The prevalence of Goldenhar-associated conditions in 668 consecutive patients with congenital deformities of the spine was 2% (14 patients).
- Failures of vertebral segmentation were the most frequent abnormality in the cervical spine, whereas failures of vertebral formation most commonly occurred in the thoracic or thoracolumbar spine.
- Spinal surgery was required in 8 patients to control the deformity.

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Infantile developmental thoracolumbar kyphosis with segmental subluxation of the spine

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We report five children who presented at the mean age of 1.5 years (1.1 to 1.9) with a progressive thoracolumbar kyphosis associated with segmental instability and subluxation of the spine at the level above an anteriorly-wedged hypoplastic vertebra at L1 or L2. The spinal deformity appeared to be developmental and not congenital in origin. The anterior wedging of the vertebra may have been secondary to localised segmental instability and subsequent kyphotic deformity.

We suggest the term 'infantile developmental thoracolumbar kyphosis with segmental subluxation of the spine' to differentiate this type of deformity from congenital displacement of the spine in which the congenital vertebral anomaly does not resolve. Infantile developmental kyphosis with segmental subluxation of the spine, if progressive, may carry the risk of neurological compromise. In all of our patients the kyphotic deformity progressed over a period of three months and all were treated by localised posterior spinal fusion. At a mean follow-up of 6.6 years (5.0 to 9.0), gradual correction of the kyphosis was seen on serial radiographs as well as reconstitution of the hypoplastic wedged vertebra to normality. Exploration of the arthrodesis was necessary at nine months in one patient who developed a pseudarthrosis.

Before they begin to walk normal infants have a long, smooth kyphotic posture from the occiput to the sacrum.¹ Once they gain head and neck control they slowly develop a cervical lordosis. The normal lumbar lordosis does not appear until the child is able to stand and walk. The spine at the thoracolumbar junction is flat in the sagittal plane.¹

The development of a thoracolumbar kyphosis in walking infants and young children may be due to spinal injury, infection, neoplasm, skeletal dysplasia, or a congenital vertebral anomaly.² In non-walking infants and young children, the primary cause of a thoracolumbar kyphosis is a myelomeningocele usually with a high-level neurological deficit.³

Most patients with a congenital kyphosis or kyphoscoliosis have a well-aligned spinal canal.⁴ In a small group with congenital kyphosis due to an anterior failure of vertebral body formation, the spinal canal is not only sharply angulated but also subluxed or dislocated at the apex of the deformity.⁵⁻⁷ Segmental subluxation or dislocation of the spine has been described as potentially the most serious form of congenital kyphosis, even though not all the patients included in the initial

reports had associated congenital vertebral abnormalities.^{5,6}

Campos et al² reported seven otherwise apparently normal children who presented with what they described as an infantile thoracolumbar kyphosis associated with single-level hypoplasia of the anterior aspect of the body of a lumbar vertebra. They noted that with time there was a spontaneous gradual correction of the kyphosis, raising the question of the effect of mechanical overload on the vertebral body and its resolution as the child developed an erect posture.

We report five children with a progressive thoracolumbar kyphosis and segmental subluxation of the spine occurring above a hypoplastic, anteriorly-wedged vertebra. These patients were thought initially to represent a variant of congenital kyphosis with displacement in the sagittal plane. However, results of surgical treatment suggest that this group of patients should not be classified as having a congenital kyphosis because the apparent congenital vertebral anomaly resolved to normality. This may represent a progressive type of infantile developmental kyphosis with associated segmental vertebral subluxation occurring at the junction between the rigid thoracic

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and more mobile lumbar spine, thereby causing secondary anterior wedging of the vertebra.

Patients and Methods

We reviewed the medical records and spinal radiographs of 750 consecutive patients who were diagnosed with a congenital spinal deformity at our institution between 1960 and 2008. Of these, 589 (79%) had a scoliosis, 106 (14%) a kyphoscoliosis and 55 (7%) a pure kyphosis. Patients who had myelomeningocele, Scheuermann's disease, neurofibromatosis, skeletal dysplasia, infection, trauma or a neoplasm were excluded, as the aim was to identify patients with congenital vertebral displacement.

We identified five boys who had a thoracolumbar kyphosis with associated vertebral displacement and a 'step-off' deformity occurring above a hypoplastic anteriorly-wedged vertebra. The vertebral anomalies were detected on anteroposterior and lateral radiographs. The degree of spinal deformity in the coronal and sagittal planes was measured using the modified Cobb method on serial spinal radiographs taken with the patient standing.^{8,9} The Meyerding method was used to measure the degree of vertebral subluxation in the sagittal plane.¹⁰ Care was taken to measure all the curves using the same anatomical landmarks on the serial spinal radiographs taken before the surgery and during follow-up.

All five patients had a whole spinal MR scan, as well as cardiac and renal assessment. Their mean age at presentation was 1.5 years (1.1 to 1.9). They were referred because the parents had noted a prominence in their child's spine. All five were neurologically normal and walked independently. None complained of back pain. On clinical examination, there was a considerable thoracolumbar kyphotic deformity, which partially corrected in the supine position.

On radiological examination, the level of the vertebral subluxation was at T12-L1 in four patients and at L1-2 in one. The anterior subluxation and 'step-off' deformity occurred only in the sagittal plane and was at the level above an anomalous vertebra (Figs 1 and 2). The anomalous vertebra at L1 (four patients) and L2 (one patient) was hypoplastic with anterior wedging and an indentation in the anterosuperior aspect of the vertebral body producing a beaked appearance. The posterior elements were normal. The wedged vertebra did not sublux posteriorly into the spinal canal and the spine was well aligned both above and below to the level of the displacement. There was no rotatory or transverse displacement and no evidence of scoliosis above or below the kyphotic segment.

MRI showed a normal appearance of the pedicles and facet joints bilaterally at the level of the anomalous vertebra in all five patients. Because of the young age of the patients and poor compliance, a supine lateral spinal radiograph in hyperextension against a bolster to assess flexibility of the deformity was not taken. However, MRI showed the kyphosis and segmental vertebral displacement to be flexible and partially correctable in the supine position (Figs 1 and 2). There were no intraspinal anomalies.

Cardiac and renal investigations were normal. None of the patients had Sprengel's shoulder, Klippel-Feil syndrome or anomalies of the ribs and chest wall. There were no medical comorbidities.

Operative treatment. The indication for surgical treatment was progression of the kyphotic deformity of 10° to 15° in all five patients over a period of three months between the initial diagnosis and decision to operate. As they were all walking, there was concern that compression of the spinal cord could develop at the site of displacement.

All five patients were treated by a localised posterior spinal arthrodesis using allograft bone without instrumentation, extending two disc levels above and below the anomalous vertebra. The patients were considered too young to have donor bone removed from their iliac crest, which otherwise would have been preferable. At operation, the posterior elements of the wedged vertebra and the facet joints above and below were found to be normal. A moulded underarm spinal jacket was applied post-operatively, with the patient under general anaesthesia and lying supine on a plaster frame. The spinal jacket was maintained for three months.

Results

Pre-operatively, the mean kyphosis was 45° (35° to 50°) which was reduced post-operatively after application of the spinal jacket to a mean of 24° (10° to 32°). However, the mean percentage segmental vertebral subluxation and step-off deformity remained unchanged at 52% (30% to 70%).

Post-operatively, all five patients were followed by serial spinal radiographs with the child standing. In one patient, after removal of the plaster jacket and over a period of six months, there was progression of the kyphosis from 32° to 43°, but with no change in the degree of vertebral displacement, which remained at 70%. Exploration nine months after the initial operation showed a pseudarthrosis, which was treated by regrafting using autologous rib graft. Repeat exploration of the fusion mass six months later confirmed solid fusion.

At the latest follow-up, at a mean of 6.6 years (5.0 to 9.0) after the initial operation, none of the children had a clinical deformity or spinal symptoms. They were all taking part in normal activities, including sport. Serial radiographs taken during the follow-up period showed gradual correction of the segmental kyphosis and vertebral subluxation as well as reconstitution of the previously hypoplastic vertebral body to normality. This occurred mainly in the first two years (Figs 1 and 2). In all patients the spine showed normal coronal and sagittal balance.

Discussion

Our patients were thought initially to have a congenital kyphosis because of partial anterior failure of vertebral formation with an associated segmental displacement of the spine and malalignment of the spinal canal with a step-off deformity in the sagittal plane. The initial diagnosis was



Fig. 1a

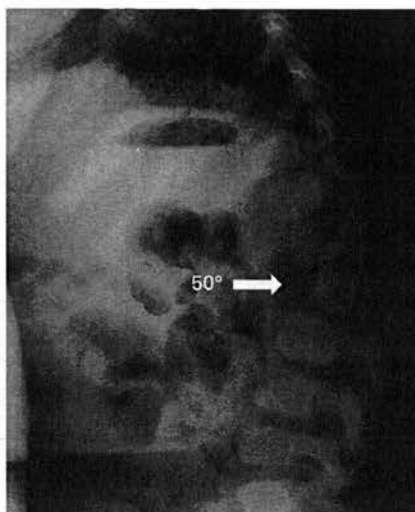


Fig. 1b



Fig. 1c

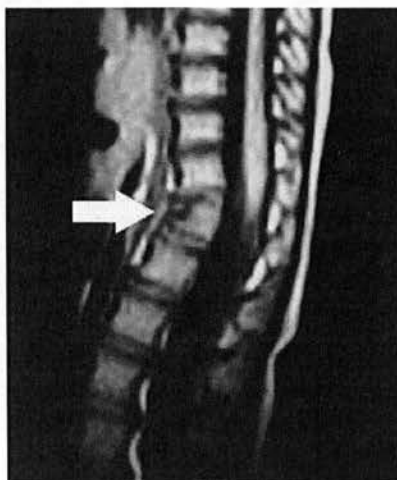


Fig. 1d

Figure 1a - Pre-operative anteroposterior radiograph of the spine at the age of 1 year 2 months showing normal spinal balance. Figure 1b - Pre-operative lateral radiograph of the spine showing a hypoplastic anteriorly wedged vertebra at L2 with anterior vertebral subluxation at the disc level above and an associated kyphosis of 50°. Figure 1c - Lateral radiograph of the spine at follow-up aged 4 years 7 months showing normal sagittal balance of the spine and complete reconstitution of the anomalous vertebra to normal. Figure 1d - Pre-operative MRI scan of the spine showing the segmental kyphosis, which is partially correctable in the supine position, as well as the hypoplastic vertebra (white arrow). Note the significant difference on the sagittal alignment of the thoracolumbar/lumbar spine between the sitting lateral radiograph (b) and the supine MRI scan (d), which confirms the presence of severe segmental instability.

based on the radiological appearance of the anomalous, anteriorly-wedged and beaked vertebra which resembled a congenital posterior wedged vertebra. This was thought to have resulted in segmental instability and subluxation of the

spine at the level above the hypoplastic vertebra, which was at the apex of the kyphosis. All our patients had normal neurological function with an intact posterior vertebral arch and normal facet joints. Despite the beaked appearance of



Fig. 2a

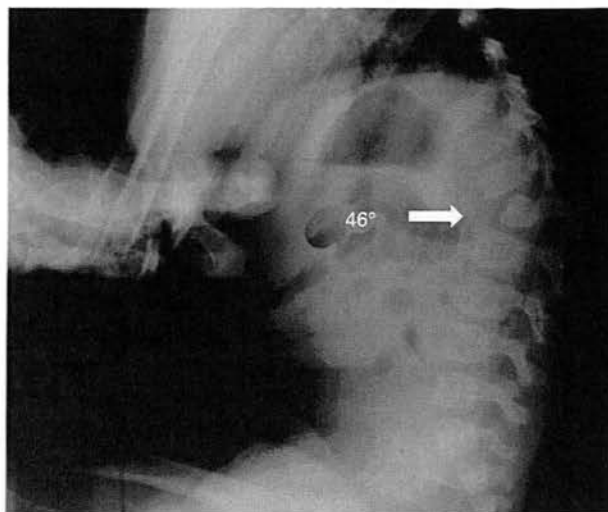


Fig. 2b



Fig. 2c

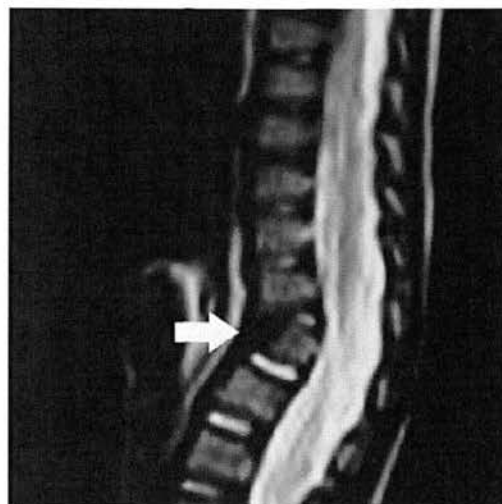


Fig. 2d

Pre-operative radiograph of the spine at the age of 1 year 3 months showing (a) a normal spinal balance, (b) a hypoplastic anteriorly-wedged vertebra at L1 with anterior vertebral subluxation occurring at the T12-L1 level and an associated thoracolumbar kyphosis measuring 46°, (c) lateral radiograph of the spine at follow-up at the age of seven years five months showing normal sagittal balance of the spine and complete reconstitution of the anomalous vertebra and (d) Pre-operative MR scan of the spine showing the segmental kyphosis, which is partially correctable in the supine position as well as the hypoplastic vertebra (white arrow). There is a considerable difference in the sagittal alignment of the thoracolumbar spine between the sitting lateral radiograph (b) and the supine MR scan (d) which confirms the presence of severe segmental instability.

the abnormal vertebra at the apex of the kyphosis, which can also be seen in patients with achondroplasia,^{11,12} there were no other clinical or radiological signs suggestive of skeletal dysplasia. MRI showed no intraspinal anomalies.

Congenital kyphosis and kyphoscoliosis is an uncommon deformity in which there is an abnormal posterior convex

angulation of a segment of the spine due to developmental vertebral anomalies that impair longitudinal growth anterior or anterolateral to the transverse axis of vertebral rotation in the sagittal plane.^{9,13} These vertebral anomalies may be due to either a defect of formation or segmentation of one or more vertebral bodies occurring anteriorly or anterolaterally.

Dubousset⁴ classified anterior defects of vertebral body formation resulting in a kyphosis or kyphoscoliosis into two types depending on the alignment of the spinal canal. In type 1 there is a partial failure of vertebral body formation with a well-aligned spinal canal and an intact posterior arch. In type 2 there is a dislocated canal which produces a step-off deformity in the sagittal plane and a bayonet-type appearance in the coronal plane, as well as abnormal posterior elements. The loss of continuity of the posterior margins of the adjacent vertebral bodies at the apex of the kyphosis has been designated as the 'step-off' sign which is the hallmark of a congenitally dislocated spine.^{4,6} However, in their original reports of rotatory dislocation of the spine, Duval-Beaupere and Dubousset⁵ and Dubousset et al⁶ included seven patients with skeletal dysplasias, five with neurofibromatosis, one with idiopathic scoliosis and only five with congenital vertebral anomalies.

In 1993, Shapiro and Herring⁷ used the term 'congenital vertebral displacement' to describe ten patients with a congenital kyphosis and an abrupt displacement of the spinal canal occurring above an anomalous vertebra which was at the apex of the deformity. They differentiated two patterns of deformity. Type A was more common and consisted of vertebral displacement occurring only in the sagittal plane as a result of a posterior hemivertebra. Type B affected all three planes and resulted in a rotatory, transverse and sagittal displacement, very similar to that of congenital dislocation of the spine as described by Dubousset et al.⁶

We thought initially that the vertebral abnormality and type of deformity in our patients represented a variant of type-A vertebral displacement.³ However, there were significant differences. With type-A displacement, the anomalous vertebra is a posterior hemivertebra and the deformity is potentially unstable because of a deficient posterior neural arch at the level of the abnormal vertebra and a tendency for the posterior hemivertebra to displace backwards into the spinal canal. This can result in either an acute paralysis after minor trauma or a gradually developing paraparesis as the deformity progresses. Also, intraspinal or systemic anomalies may be present.

In our group of patients with an angular kyphosis, the displacement also occurred in the sagittal plane above an anteriorly-wedged vertebra with a 'step-off' deformity. However, the spine was well aligned above and below the level of subluxation and the hypoplastic anteriorly-wedged vertebra was stable with no displacement posteriorly into the spinal canal. The posterior vertebral elements and pedicles were normal and we did not find any intraspinal abnormalities. All our patients showed progression of their deformity before operative treatment and although they had normal neurology there was concern that increasing deformity could cause compression of the spinal cord and neurological complications.

We found that early posterior spinal arthrodesis, in the presence of continuing anterior vertebral growth, allowed not only gradual spontaneous correction of the kyphosis

but also complete reduction of the associated vertebral subluxation (Figs 1 and 2). There was also complete reconstitution of the anomalous wedged vertebra to normality in all five patients which occurred mainly in the first two years after posterior spinal arthrodesis. This finding suggests that the hypoplasia and anterior wedging of the vertebral body could be developmental in origin as a result of secondary disturbance of the anterior growth of the affected vertebra at the level of mechanical instability. The favourable clinical outcome after surgery may be an example of the Hueter-Volkman principle in which the growth plates have increased growth in response to tension produced by the posterior fusion.¹⁴

Kim and Weinstein¹⁵ described two patients with an atypical form of congenital kyphosis with associated segmental vertebral displacement. The appearance of the anteriorly hypoplastic vertebra in their report was similar to that in our patients. However, it differed in that their patients had short pedicles and segmental narrowing of the spinal canal at the level of the displacement suggestive of a congenital abnormality. They performed a short anterior and a longer posterior spinal arthrodesis across the apex of the kyphosis. The kyphotic deformity in these two patients took much longer to correct when compared with our study. This may have been due to the short anterior spinal fusion limiting the potential for spontaneous anterior correction, which could only occur in the unfused levels above and below those which were included in the longer posterior fusion.

The absence of other abnormalities in the formation of the vertebral column and the neural structures apart from the anterior wedging of the hypoplastic vertebra differentiates the type of anomaly and associated deformity seen in our patients from congenital displacement of the spine. We therefore suggest that the term 'infantile developmental kyphosis with segmental subluxation of the spine' should be used to describe the condition seen in our five patients.

One of our patients developed a pseudarthrosis after posterior spinal arthrodesis with further progression of the kyphosis. Unfortunately, in young children it is not possible to assess the quality of a posterior spinal fusion simply on plain radiography. We therefore recommend exploration of the fusion mass if there is any loss of correction after removal of the spinal jacket following the initial arthrodesis to either repair a pseudarthrosis or augment a weak spinal fusion with additional autologous bone graft. In our patient, the pseudarthrosis was explored and repaired at nine months when the kyphosis had relapsed to 43°. The patient went on to develop a normal sagittal profile five years after the initial spinal procedure. Long-term follow-up to skeletal maturity will be required, even though surgery has been successful to date in all our patients.

Campos et al² described seven infants with normal neurological function who developed a thoracolumbar kyphosis which was thought to have occurred secondary to lumbar hypoplasia affecting the L1 or L2 level. The appear-

ance of the anomalous vertebra in their report was similar to that seen in our patients. The authors did not comment on the presence of segmental subluxation occurring at the level above the anteriorly wedge-shaped vertebra. However, Figure 1 in their article shows evidence of such subluxation.⁴ The mean age at presentation was 5.3 months (birth to 11 months) and the mean angle of the kyphosis in sitting radiographs was 34.2° (24° to 41°) at a mean age of 12 months. The mean kyphosis shown in the first standing radiograph was 19.7° (10° to 27°). All the patients were treated by observation and the kyphosis gradually improved spontaneously. At a mean follow-up of 5.7 years, all the patients were asymptomatic and the radiological evaluation showed correction of the kyphosis to nearly normal.

The difference between these patients and our group is that all our patients presented later when they had started walking with a more severe and progressive thoracolumbar kyphosis, as well as segmental subluxation of the spine at the level above the anteriorly-wedged vertebra which we felt warranted surgical treatment. These two groups of patients may represent a spectrum of the same deformity in which some resolve spontaneously whereas others progress and require treatment. It is interesting that in both groups the kyphosis developed at the transitional thoracolumbar junction which suggests a mechanical cause for the condition. One weakness of our study is that in the presence of rapid kyphosis progression none of our patients were initially managed non-operatively. Because of the developmental nature of the deformity serial casting or bracing might have been effective.

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Guest Speaker

Spinal Growth and Congenital Deformity of the Spine

Michael J. McMaster, MD, FRCS

Ladies and gentleman, I am honored to have been invited by the Scoliosis Research Society to give the Harrington Lecture. We are all aware that congenital deformities of the spine are caused by the presence of developmental vertebral anomalies such as this hemivertebra, which produce a localized imbalance of the longitudinal growth of the spine. However, the term “congenital” is slightly misleading because it implies that the curvature is apparent at birth, but this is not necessarily so. It is the vertebral anomalies that are present at birth, and the clinical deformity only develops with spinal growth and may not become apparent until later childhood when the diagnosis is made radiographically.

In a personal study of 670 patients with congenital spine deformities seen at my Edinburgh clinic, I found that 80% had a scoliosis, 14% had a kyphoscoliosis, and 6% had a pure kyphosis. It should be appreciated that these deformities are not separate entities but are part of a gradually blending spectrum of deformities. The type of deformity that develops depends on whether the impaired growth occurs unilaterally in the coronal plane producing a scoliosis, or occurs anterior or anterolateral to the transverse axis of vertebral rotation in the sagittal plane producing a kyphosis or kyphoscoliosis.

In the normal spine, growth occurs at the endplates on the upper and lower surfaces of the vertebral bodies. This growth occurs symmetrically, and the spine remains balanced in the coronal and sagittal planes. In the presence of a congenital vertebral anomaly, there is an absence or deficiency in the number of growth plates on one side of the spine, resulting in a localized unilateral longitudinal growth imbalance and an increasing spinal curvature as the child grows. The greater the growth imbalance, the more severe the deformity, and this only stabilizes at skeletal maturity when the growth plates fuse. However, the growth velocity of the spine is not uniform, and there are 2 periods of accelerated growth. The first occurs up to the age of 3 years and the second during the adolescent growth spurt, which usually occurs in girls between the ages of 10 and 13 years, and in boys 2 years later. In the intervening period, there is steady linear growth.

If we look at the age at diagnosis of patients with a congenital spine deformity presenting at my clinic, we see that the majority were diagnosed during these periods of rapid spinal growth, when the shape of the back was changing most rapidly and the deformity became more obvious. All degrees of severity of scoliosis were seen at all ages. Some patients presented with small curves that progressed minimally, whereas others progressed rapidly to become an extreme deformity at an early age.

The skill in managing a patient with a congenital spine deformity lies not just in the ability to perform major complex surgery at a late stage but primarily in recognizing those curves with a severe spinal growth imbalance at an early age and applying prophylactic surgical treatment to prevent curve progression. It is much better to treat a patient at an early stage when the curve is small rather than to wait until a severe rigid deformity has developed, when it is necessary to perform potentially dangerous surgical salvage procedures. This will be the main topic of my lecture.

To understand the natural history of congenital spine deformities and the great disparity in prognosis, it is necessary to correlate the principles of normal growth of the spine with the pathologic anatomy and growth imbalance produced by the various types of congenital vertebral anomaly that may produce a scoliosis, kyphosis, or kyphoscoliosis. The development of the vertebral column is a very rapid process, and the complete anatomic pattern is formed in mesenchyme during the first 6 weeks of intrauterine life. The vertebral anomalies that cause a congenital scoliosis develop during this period, and once the mesenchymal mold is established, the cartilaginous and bony stages follow that pattern, and the anomalies are fully established at birth.

The classification of congenital scoliosis is based on this embryologic mal-development of the spine. There are 2 groups of vertebral anomalies that can produce a scoliosis.¹ First, those caused by defect of vertebral formation. The most common of which is a hemivertebra, occurring in 38% of my patients. Second, there are those caused by a unilateral defect of segmentation of ≥ 2 vertebrae. This resulted in a unilateral unsegmented bar in 30% and a unilateral unsegmented bar with contralateral hemivertebrae in 12% of patients. An additional 12% of patients could not be classified because they had a complex mixture of anomalies. Wedged and block vertebrae are not often seen by themselves because the resulting deformity is so small that it is not usually recognized. These anomalies are much more commonly seen as part of a complex of mixed vertebral anomalies that

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cannot be specifically diagnosed and may produce a more severe deformity.

A hemivertebra is the most common cause of a congenital scoliosis, but the severity of the deformity varies greatly, and there is debate as to the necessity and timing of treatment.² Opinions range through the spectrum from total excision to total neglect of the hemivertebra. To rationalize this disparity, it is necessary to have an understanding of the growth of the spine, and the problems associated with the different types and sites of hemivertebrae.

The potential for a hemivertebra to cause a significant spine deformity depends on 3 factors. The first and most important factor is the pathologic anatomy and relationship of the hemivertebra to the adjacent hemivertebrae in the spine. The hemivertebra may be fully segmented, which is most common, semi-segmented, or incarcerated, which is least common. It is important to distinguish between these 3 types because each has a different growth potential, and the severity of the resulting scoliosis is related to the degree of segmentation. Second, the site of the hemivertebra is important, especially those occurring at the lumbosacral junction. The third factor is the number of hemivertebrae and their relationship to each other in the spine. Is there a single hemivertebra, 2 hemivertebrae, are they on the same side, or are they opposing?

Let us look at a fully segmented hemivertebra. It has a normal disc above and below, and is completely separate from its adjacent vertebrae. There is an absence of 2 growth plates on the unformed side, whereas relatively normal growth occurs on the upper and lower surfaces of the hemivertebra. As the hemivertebra grows, it acts as an enlarging wedge, resulting in an increasing scoliosis. This type of hemivertebra may occur anywhere in the spine with a significant number at the lumbosacral junction, where they can cause a major problem. Without treatment, progression of a scoliosis caused by a single fully segmented hemivertebra occurring in the thoracic and lumbar regions can be difficult to predict and requires careful monitoring. However, the majority of curves progress relatively slowly at 1° or 2° per year. The most pernicious and deforming type of hemivertebra occurs at the lumbosacral junction. Here, it causes an oblique takeoff of the lumbar spine from the sacrum, and the patient lists to one side. A compensatory curve develops above, but this is never sufficient to balance the spine. Initially, this compensatory curve is mobile and correctable but, with time, becomes fixed and rotated, and is the major deformity. The time to treat these patients is in the first few years of life, before the compensatory curve becomes fixed and not at this late stage.

Two unilateral hemivertebrae that are less common but have a much worse prognosis. Here, there is an absence of 4 growth plates on one side of the spine, resulting in a much higher growth imbalance. These curves usually progress at 3° to 4° per year, and the majority will exceed 50° by the age of 10 years. By skeletal maturity,

the majority of curves will be more than 70°. All these patients require prophylactic treatment to balance the growth of the spine at an early stage.

Two opposing hemivertebrae have a more variable prognosis depending on the type of hemivertebra and whether the hemivertebrae are close together or more widely separated in different regions of the spine. If the hemivertebrae are close together in the same region, they tend to balance each other and cause only a minimal cosmetic deformity, and no treatment is required. However, if the hemivertebrae are more widely separated in different regions of the spine, the curves are often unbalanced, causing spinal decompensation, which is much more deforming.

Not all hemivertebrae are the same. A semi-segmented hemivertebra is synostosed to its neighboring vertebra and has only 1 disc space either above or below. As a result, 2 growth plates are obliterated on the convexity, and this tends to balance the absence of the 2 growth plates on the unformed side of the hemivertebrae. Although growth of the spine is theoretically balanced, the hemivertebra can cause a tilting of the spine and induce a slowly progressive scoliosis, but this does not usually require treatment. This type of hemivertebra mainly occurs in the lumbar region, often at the lumbosacral junction.

An incarcerated hemivertebra is a small ovoid piece of bone lying in a niche in the spine, which remains straight. The disc spaces above and below the hemivertebra are usually narrow and poorly formed, indicating a poor growth potential. This type of hemivertebra usually occurs in the thoracic region. The resulting scoliosis deteriorates very slowly, if at all, and rarely exceeds 20° at skeletal maturity. This does not require treatment.

When treating a patient with a congenital scoliosis caused by a hemivertebra, it is important to appreciate that it is not possible to create growth on the side of the spine that is not growing. It is only possible to balance growth by retarding growth on the convexity. This retardation can be achieved by either a convex growth arrest procedure or excising the hemivertebra. A convex growth arrest procedure tethers the growth on the convexity, with the objective of allowing continuing growth on the concavity to correct slowly the deformity. Excision of the hemivertebra removes the primary cause of the scoliosis, which is the enlarging wedge on the convexity at the apex of the deformity. For either of these procedures to be effective, they should be performed before the age of 5 years, while the deformity is still small and the compensatory curves are mobile.

A unilateral unsegmented bar is the second most common cause of congenital scoliosis. Here, there is a unilateral failure of segmentation of ≥ 2 vertebrae. The unsegmented bar does not contain growth plates and, therefore, does not grow longitudinally, whereas some degree of growth occurs on the convexity, resulting in a progressive scoliosis.

The unsegmented bar may occur anywhere in the spine from the upper thoracic region to the sacrum, and

no one region is more commonly affected than the other. The mean length of the unsegmented bar is over 3 vertebrae (range 2–8). One would assume that the more extensive the bar, the more severe the rate of progression and final severity of the scoliosis. However, I found that there was not always a direct relationship between the length of the bar and rate of deterioration of the scoliosis. Of much more importance is the growth potential on the convexity of the curve. Here we see a small curve, caused by a short unsegmented bar, but the disc spaces on the convexity are widely open, indicating good growth potential, and this produces a large curve at an early age. It is the unbalanced growth of the spine on the convexity that drives the deformity and not the unsegmented bar. If there was no convex growth, there would be no deformity.

Here is a similar small curve caused by a short unsegmented bar, but in this case, the disc spaces on the convexity are narrow, indicating poor growth potential, and a much smaller curve develops. Therefore, it is important when assessing prognosis to look at both sides of the spine and not just at the extent of the unsegmented bar. The age of the patient and onset of the adolescent growth spurt also have an effect, with much more rapid deterioration occurring during puberty. Without treatment, the majority of patients with a unilateral unsegmented bar have a bad prognosis, with the scoliosis exceeding 50° by the age of 10 years and a severe rigid deformity. These patients require early prophylactic treatment to balance the growth of the spine at an early age.

We know that a congenital scoliosis caused by a unilateral unsegmented bar has a bad prognosis. However, there is a smaller, less well-recognized group of patients who have an even worse prognosis. These patients have not only a unilateral unsegmented bar, but also ≥ 1 hemivertebrae on the contralateral side at the same level.³ These hemivertebrae produce an even greater growth imbalance than if there had been an unsegmented bar alone. If left untreated, this can result in some of the most rapidly progressive and severe deformities seen by the spine surgeon.

These vertebral anomalies occur in all regions of the spine, with the mean length of the unsegmented bar extending over 5 vertebrae and a mean of 3 contralateral hemivertebrae (range 1–8 vertebrae). The diagnosis of this type of anomaly is not always easy. Radiographically, the vertebral anomalies are most clearly seen in the first few years of life, but as the curve deteriorates, the hemivertebrae often become obscured by the severity of the deformity. The crankshaft effect produced by the continuing growth of the hemivertebrae anterolaterally combined with the tethering effect of the unsegmented bar on the concavity results in severe vertebral rotation and distortion of the rib cage. Without treatment, these curves will progress on average 6° per year, and the majority will exceed 50° by 2 years of age. The chest deformity that usually occurs at an early age can impair lung growth and development.

There is also an important secondary feature, common to all types of congenital scoliosis, relating to the site of the vertebral anomalies that contributes significantly to the overall disability and deformity of the patient. A congenital mid-thoracic curve with its apex at T5, T6, or T7, especially those caused by a unilateral unsegmented bar with or without contralateral hemivertebrae, is frequently associated with the development of a long secondary structural curve in the lower thoracic or thoracolumbar regions. As the congenital curve deteriorates with growth, it produces a rotational torque that is transmitted further down the spine, resulting in a secondary scoliosis to the opposite side. This secondary curve does not contain any congenital anomalies, and is initially compensatory and correctable. However, it often deteriorates more rapidly than the congenital curve, and later becomes fixed and severely rotated, producing the major cosmetic deformity and requiring treatment.

Congenital rib and chest wall anomalies commonly occur on the concavity of a thoracic or thoracolumbar scoliosis caused by a unilateral unsegmented bar, with or without contralateral hemivertebrae.⁴ These rib anomalies may have a unilateral tethering effect on the spine, but this is of minor significance in comparison with the main driving force for the spinal deformity, which is the severe growth imbalance produced by the vertebral anomalies.

So far, I have spoken only of a congenital scoliosis. A congenital kyphosis and kyphoscoliosis are much less common than a congenital scoliosis but are even more important to understand because of a potential complication that occurs with spinal growth and does not affect a congenital scoliosis. Of the 136 patients with a congenital kyphosis or kyphoscoliosis seen at my Edinburgh clinic, 12 who were previously neurologically normal presented with a progressive neurologic deficit caused by spinal cord compression.

A congenital kyphosis or kyphoscoliosis is an abnormal posterior convex angulation of a segment of the spine caused by developmental vertebral anomalies, which impair anterior longitudinal growth in the sagittal plane. The anterior longitudinal growth imbalance is often asymmetric, producing an associated scoliosis in the same region, but the major deformity is the kyphosis. The classification of the various types of congenital kyphosis and kyphoscoliosis is similar to that for a congenital scoliosis and is based on the embryologic maldevelopment of the spine but occurring at a later stage of intrauterine development. Of my patients, 65% had an anterior failure of vertebral body formation, 20% had an anterior failure of vertebral body segmentation, 10% had mixed anomalies, and 5% could not be classified. The resulting deformity occurred at all levels of the spine but was most common at the thoracolumbar junction, and less frequent in the upper thoracic and lower lumbar regions. However, all degrees of severity of deformity occurred at all levels of the spine.

An anterior failure of vertebral body segmentation is thought to be caused by bony metaplasia in the anterior part of the anulus fibrosis and ring apophysis, occurring during the late chondrification and ossification stages of embryologic development. This results in an anterior unsegmented bar, which contains no growth plates. However, some degree of growth occurs posteriorly in the growth plates above and below the residual discs and in the neural arch, resulting in an increasing kyphosis. These patients usually have a pure kyphosis with very little, if any, associated scoliosis, and the deformity deteriorates relatively slow at less than 2° per year and does not result in spinal cord compression. A few patients with an anterior unsegmented bar may have a larger deformity, but this is usually present from an early age, and does not progress significantly and does not result in spinal cord compression, regardless of the size of the kyphosis.

An anterior failure of vertebral body formation is the most common cause of a congenital kyphosis or kyphoscoliosis and can result in spinal cord compression. This type of vertebral anomaly is thought to be caused by a partial failure of vascularization of the developing cartilaginous centrum, resulting in a deficiency in the developing vertebral body. Growth occurs in the discs above and below the residual vertebra and in the posterior elements, but there is an absence of 2 growth plates anteriorly. There are 4 patterns of vertebral body malformation: the most common of which is a posterolateral quadrant vertebra; followed by a butterfly vertebra; a posterior hemivertebra; and, finally, an anterior wedged vertebra. The rate of progression and severity of the curvature is proportional to the severity of the anterior failure of vertebral body formation and usually produces a kyphoscoliosis. A posterolateral quadrant vertebra and posterior hemivertebra have the highest anterior deficiency and, therefore, produce the most severe deformity.

Here is an example of a posterolateral quadrant vertebra at the thoracolumbar junction. This type of kyphosis deteriorates much more rapidly than that caused by an anterior unsegmented bar. Without treatment, the majority of these children will have a kyphosis of more than 80° by the age of 10 years, and there is a significant risk of spinal cord compression. All require early prophylactic treatment with a localized posterior spine fusion to balance the growth of the spine before the child is 5 years old and the kyphosis exceeds 40°. This procedure provides a posterior tether and the opportunity for the kyphosis to correct with continuing anterior growth, and eliminates the risk of spinal cord compression.⁵

Spinal cord compression is the most dreaded complication of a congenital kyphosis or kyphoscoliosis but only occurs in patients with an anterior failure of vertebral body formation. The reason for this is that these anomalies are unstable and tend to extrude backwards into the spinal canal at the apex of the angular deformity as the kyphosis deteriorates with spinal growth. The vast

majority of patients with a congenital kyphosis or kyphoscoliosis have their curve apex and vertebral anomalies in the thoracolumbar region. However, the greatest risk of spinal cord compression occurs when the curve apex is in the mid and upper thoracic regions. Here, the segmental blood supply to the spinal cord is poorest, the spinal canal is narrowest, and the cord is least mobile.

The frequency of spinal cord compression is difficult to estimate because many patients are now treated with prophylactic surgery at an early age. In the 2 largest reported series,^{6,7} spinal cord compression developed in 18% of patients with an anterior failure of vertebral body formation. However, the true frequency if all patients were untreated would be much greater. The onset of spinal cord compression can occur at any age but is most frequent during the adolescent growth spurt. The size of the kyphosis at the onset of paralysis varies. Paraparesis developed in 1 of my patients with only a 60° kyphosis, whereas others with a kyphosis of more than 100° remained neurologically intact. However, all patients who have a neurologic deficit developing as a result of spinal cord compression will always have progression to paraplegia if left untreated.

In conclusion, the key to successful treatment of all types of congenital spine deformity, whether it is a scoliosis, kyphosis, or kyphoscoliosis, depends on 3 principles. First, early diagnosis before the child is 5 years old and the curve, whether it is a scoliosis or kyphosis, exceeds 40° and the compensatory curves become fixed. Second, the ability to anticipate what is likely to happen based on the type and sites of the vertebral anomalies, as well as the age of the patient and amount of the spinal growth remaining. Finally, the application of prophylactic surgical treatment to balance the growth of the spine at an early age and before the development of a significant deformity. Ladies and gentleman, it has been my privilege to give the Harrington Lecture, and I look forward to seeing you at the Scoliosis Research Society Meeting in Edinburgh in 2007.

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JAMES IV LECTURE

Congenital deformities of the spine

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King James IV Lecture given at the Annual Congress of the British Orthopaedic Association, International Convention Centre, Birmingham on 12th September 2001

Congenital anomalies of the vertebrae producing a scoliosis, kyphoscoliosis or kyphosis are potentially serious conditions, which can, on occasion, result in an extremely severe rigid spinal deformity with possible spinal cord compression. The key to successful management depends on: (1) Early diagnosis while the curve is still small. (2) Anticipation of the likely prognosis based on the type and site of the vertebral anomaly, the degree of growth imbalance it produces and the amount of spinal growth remaining. (3) Preventing progression of the deformity and this may necessitate surgical treatment in the first few years of life. It is much better to carry out a relatively simple operation to balance the growth of the spine at an early stage than to wait and perform potentially hazardous anterior and posterior spinal surgery as a salvage procedure at a later stage.

Keywords: Congenital scoliosis, congenital kyphosis, natural history, surgery

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INTRODUCTION

Congenital deformities of the spine are due to the presence of developmental vertebral anomalies, which produce a localised imbalance in the longitudinal growth of the spine. The term congenital is slightly misleading because it implies that the deformity is present at birth but this is not necessarily so. It is the vertebral anomalies, which are present at birth, and the clinical deformity may not become apparent until later childhood when the diagnosis is made radiographically.

Congenital deformities of the spine are relatively uncommon. However, I have been able to study 670 patients with this type of deformity seen at my Edinburgh Spine Deformity Clinic. This is one of the largest groups of reported patients and has been the subject of a series of publications on the natural history and management of the condition.¹⁻⁶ I found that all degrees of severity of curvature were seen at all ages. Some patients were first seen in their teenage years with small curves, which progressed minimally. Others presented in the first few years of life with curves, which deteriorated rapidly, becoming extreme deformities at an early age, (Figures 1, 2 and 3) and a few developed spinal cord compression and paraplegia. These severe curves were often rigid, and surgical correction was difficult and dangerous.

The skill in managing a patient with a congenital spine deformity lies not in the ability to perform major complex salvage surgery at a late stage, when there is a severe rigid deformity, but in recognizing those curves with a bad prognosis at an early

stage and applying prophylactic surgical treatment to prevent curve progression and possible neurological complications. Planning such treatment requires a thorough knowledge of the natural history of all types of congenital spine deformity and the methods of treatment which are available.

PATHOGENESIS

In order to understand the great disparity in prognosis for congenital spine deformities it is necessary to correlate the principles of normal growth of the spine with the pathologic anatomy of the various types of developmental vertebral anomalies that may cause the deformity (Figure 4).

The embryological development of the spine is a very rapid process and the complete anatomical pattern is formed in mesenchyme during the first six weeks of intrauterine life. Once the mesenchymal mould is established the cartilaginous and bony stages follow that pattern. Vertebral anomalies appear during the mesenchymal and early chondroformation stages and are due to either a defect of vertebral formation or a defect of segmentation of the primitive vertebrae. The vertebral anomalies are fully established at birth.

The neural axis also develops during this early embryological period and it is not surprising, therefore, that neural and vertebral malformations often co-exist. In this lecture, I will deal only with 'closed' congenital deformities in which the skin overlying the spine remains intact although there may be occult intraspinal anomalies, and not with those who have associated myelomeningocele.

Normal longitudinal growth of the spine occurs at the end plates on the upper and lower surfaces of the vertebral bodies.² This growth occurs symmetrically and, as a result, the spine

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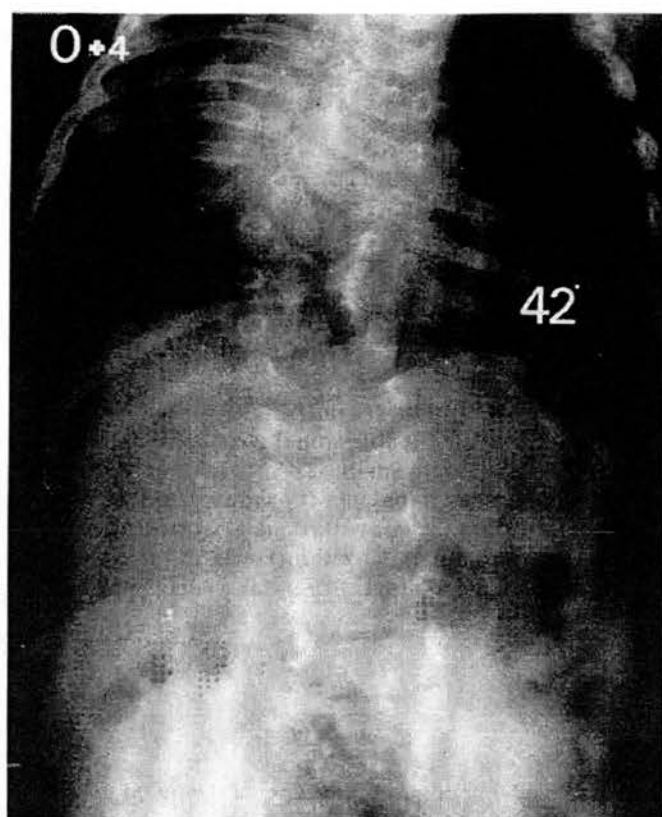


Figure 1: Infant girl aged 4 months with a thoracic scoliosis due to a unilateral unsegmented bar on the concavity with 3 adjacent contralateral hemivertebrae on the convexity at the same level. No treatment was given.

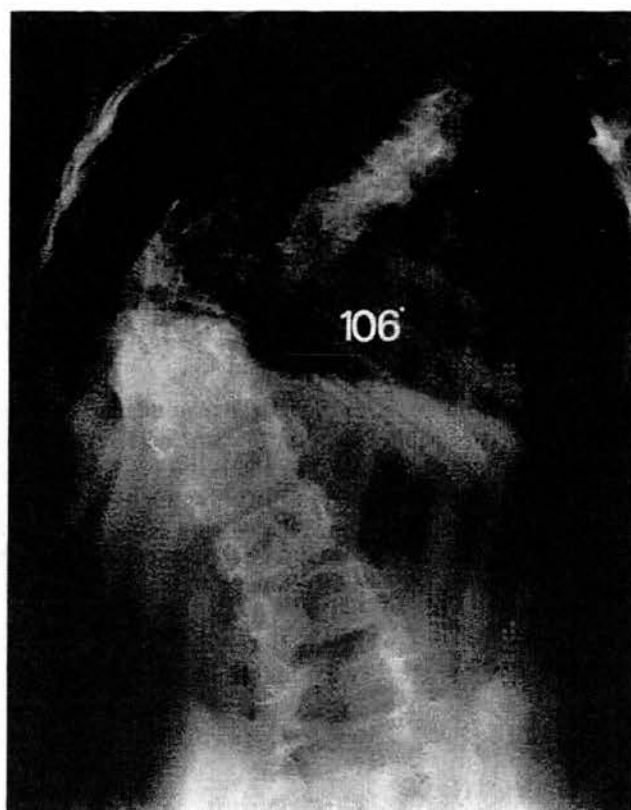


Figure 2 & 3: By the age of 9 years and 10 months the scoliosis had increased to 106 degrees and there was marked malalignment of the trunk combined with distortion of the rib cage reducing the vital capacity



Figure 3

grows in a balanced manner without a pathological curvature. However, in the presence of a congenital vertebral anomaly there is an absence or deficiency in a number of growth plates on one side of the spine, resulting in a longitudinal growth imbalance and an increasing curvature as the child grows.

The rate of deterioration and final severity of a congenital spine deformity is proportional to the degree of growth imbalance produced by the vertebral anomalies. An early onset of the deformity carries a bad prognosis because this indicates a severe growth imbalance, which will continue to skeletal maturity when the growth plates fuse. The greater the growth imbalance, the more severe the deformity. However, the rate of deterioration is not uniform and accelerates during the adolescent growth spurt, which is at its peak in girls around the age of 12 years and in boys at 14 years.

The type of spinal deformity which develops depends on whether the impaired growth occurs unilaterally, producing a pure scoliosis, or lies anterolaterally or anterior to the transverse axis of vertebral rotation in the sagittal plane, producing a kyphoscoliosis or a pure kyphosis. In my series of 670 patients, there was a gradual blending spectrum of deformities with 80% having a scoliosis, 14% kyphoscoliosis and 8% a kyphosis.

The classification of congenital spine deformities is based firstly, on the pattern of curve, ie scoliosis, kyphoscoliosis or

CONGENITAL SCOLIOSIS

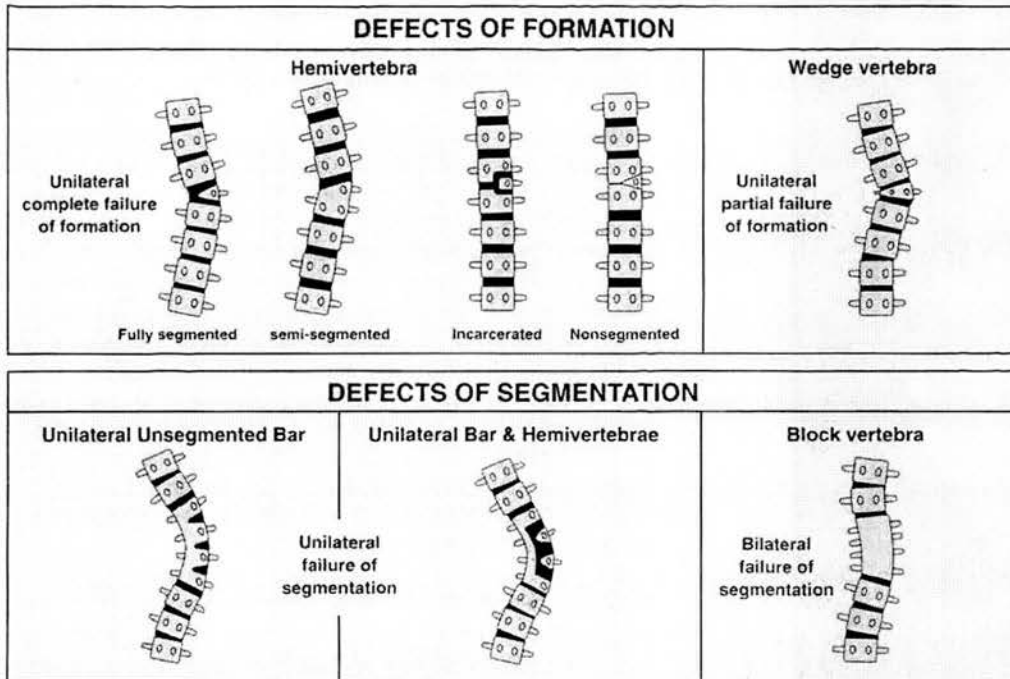


Figure 4

kyphosis, and secondly, on the type of vertebral anomaly which produced the deformity.^{1,5,8,9} There are two basic groups of anomalies; those due to a defect of vertebral formation and those caused by a defect of segmentation of one or more vertebrae. (Figure 4) There is also a smaller group with mixed or unclassifiable anomalies.

CONGENITAL SCOLIOSIS

Defect of Formation: A hemivertebra is the commonest cause of a congenital scoliosis and is due to a complete failure of a vertebra to form on one side. Longitudinal growth occurs on the upper and lower surfaces of the hemivertebra, whereas there is an absence of two growth plates on the unformed side. (Figure 4) As the hemivertebra grows, it acts as an enlarging wedge on one side of the spine resulting in an increasing scoliosis. Hemivertebrae may be single or multiple and occur at all levels of the spine.^{1,3}

Progression of a scoliosis due to a single fully segmented hemivertebra can be difficult to predict and requires careful monitoring. The majority of these curves progress relatively slowly at 1-2 degrees per year and by skeletal maturity can produce a moderately severe scoliosis of over 50 degrees in the lower thoracic and thoracolumbar regions. However, the most pernicious and deforming hemivertebra occurs at the lumbosacral junction where it causes the lumbar spine to take-off obliquely from the sacrum and the patient lists to the side producing a significant deformity. A compensatory curve develops above but this is never sufficient to balance the spine and early surgical treatment is required to prevent this deformity from developing.¹⁰

Two fully segmented unilateral hemivertebrae are less common but have a much worse prognosis because there is an

absence of four growth plates on the concavity of the curve. These curves progress much more rapidly at 3 to 4 degrees per year and the majority exceed 50 degrees by the age of 10 years. All require early prophylactic surgical treatment.

Two opposing hemivertebrae have a more variable prognosis. If they are close together they tend to balance each other, causing only two small kinks in the spine and minimal cosmetic deformity. However, if the hemivertebrae are widely separated in different regions of the spine, the resulting curves are often unbalanced causing the trunk to list to one side and prophylactic surgical treatment may be required.

Not all hemivertebrae are fully segmented. A semisegmented hemivertebra is synostosed to one of its neighbouring vertebrae and there is relatively little growth imbalance. These curves progress very slowly and no treatment is required. An incarcerated hemivertebra is a small ovoid piece of bone with very poor growth lying in a niche scalloped out of the adjacent vertebrae. In these patients the spine remains straight and no treatment is required.

In treating a congenital scoliosis due to a hemivertebra it is important to appreciate that it is not possible to create growth on the side of the spine, which is not growing. It is only possible to prevent progression by retarding growth on the convexity. In these circumstances a spinal brace is never an alternative to appropriate surgical treatment.

A convex growth arrest procedure (combined anterior and posterior convex hemiepiphyseodesis) is the optimum form of early prophylactic surgical treatment for a fully segmented hemivertebra.^{11,12} This procedure is best applied to patients younger than 5 years of age with a scoliosis less than 40



Figure 5: Infant aged 2 years with a 33 degree right thoracolumbar scoliosis due to a single fully segmented hemivertebra at L1. A combined anterior and posterior convex growth arrest procedure was performed

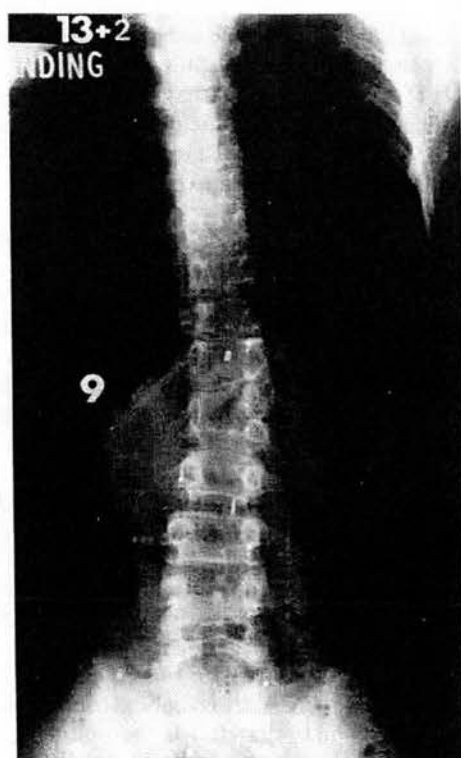


Figure 6: By the age of 13 years 2 months the curve had improved to 9 degrees

degrees. The object of the surgery is to balance the growth of the spine by preventing further unbalanced growth on the convexity at the site of the hemivertebra. Theoretically, this should allow the scoliosis to correct slowly by means of continuing growth on the concavity. (Figures 5 and 6)

Excision of a hemivertebra is theoretically attractive as a prophylactic procedure because it removes the primary cause of the scoliosis, which is the enlarging wedge on the convexity at the apex of the scoliosis.¹³⁻¹⁵ The hemivertebra is excised both anteriorly and posteriorly through the same combined two stage surgical approach as used for a convex growth arrest procedure. This creates a wedge osteotomy of the spine which, when closed, produces maximum correction and realignment of the spine. However, the majority of hemivertebrae, except for a lumbo-sacral hemivertebra, do not cause significant imbalance and are probably more safely treated by a convex growth arrest procedure, which does not require opening into the spinal canal. The best results of a hemivertebra excision are obtained when surgery is carried out at an early age before the compensatory curves become fixed.

Defect of Segmentation: A unilateral unsegmented bar is the second most common cause of a congenital scoliosis and is due to a unilateral failure of vertebral segmentation affecting two or more vertebrae. The mean length of the bar is usually over three vertebrae. The unsegmented bar does not contain any growth plates and, therefore, does not grow longitudinally, whereas some degree of growth occurs on the opposite side of the spine. The rate of progression and final severity of the scoliosis depends not only on the extent of the bar but more importantly on the growth potential on the convexity which drives the curve.¹¹ This produces a much greater deformity than a single hemivertebra. These curves progress at a mean 5 degrees per year and usually exceed 50 degrees by the age of 10 years. All of these patients require early surgical treatment, preferably before the age of 5 years.

A congenital scoliosis due to an unsegmented bar has a bad prognosis. However, there is a smaller less well recognised group of patients who have an even worse prognosis. (Figures 1, 2 and 3) These patients have not only a unilateral unsegmented bar but also one or more hemivertebrae on the contralateral side at the same level. These hemivertebrae produce an even greater growth imbalance than if there had been a unilateral unsegmented bar alone.^{4,16} The mean length of the bar is over three vertebrae with a mean two contralateral hemivertebrae. This causes the most severe and rapidly progressive of all types of congenital scoliosis and, without treatment, these curves deteriorate at 6 degrees or more per year and usually exceed 50 degrees by the age of four years. All require very early surgical treatment, as soon as the diagnosis is made.

The ideal treatment for a congenital scoliosis due to a unilateral unsegmented bar, with or without contralateral hemivertebrae, is to recognise the poor prognosis at an early stage when the curve is small and perform a spinal fusion to prevent further progression.¹⁷ A convex growth arrest procedure or simply dividing the unsegmented bar will not correct this type of deformity because there is no growth potential in the

unsegmented bar on the concavity of the curve.

The object of an early prophylactic spine fusion is to stabilise the curve at an early stage by creating a solid thick fusion that will stop the unbalanced growth of the spine. This usually necessitates a combined anterior and posterior fusion to control the severe spinal growth imbalance in these patients. The best results are achieved when the surgery is carried out before the age of two years.^{4,17} The argument that an early spine fusion will stunt the growth of the spine in these young patients is of no relevance because the unsegmented bar is not contributing to vertical height and only making the spine more crooked. There is no perfect treatment and it is much better to have a short relatively straight patient, with a fused spine, than a patient with an unfused spine, who is severely deformed and even shorter because of the severe spinal curvature.

In an older child with a moderate deformity, partial correction of the scoliosis may be attempted at the time of spine fusion but correction only occurs in the mobile segments above and below the unsegmented bar. The object of surgery is to achieve overall balance of the spine rather than excessive correction of the congenital curve which could be dangerous.^{16,17}

A magnetic resonance imaging scan of the spine is essential in all patients before intra-operative correction of the deformity is attempted using spinal instrumentation. This may reveal the presence of an intra spinal anomaly, such as a diastematomyelia, that could be tethering the cord. A diastematomyelia has been reported in 5 to 20% of patients with a congenital scoliosis and, if the spur is not removed, these patients could develop serious neurological complications due to traction on the spinal cord at the time of correction of the scoliosis.^{1,19-22} Of the various types of scoliosis, the congenital variety carries the highest risk of neurological complications after intraoperative correction.²³

Patients who present at a late stage, with a severe rigid deformity causing significant spinal imbalance or fixed pelvic obliquity, require correction by means of an anterior and posterior closing wedge osteotomy of the vertebral column combined with spinal instrumentation, to maintain correction and an extensive fusion of the whole of the curvature.⁴ A closing wedge osteotomy decreases the neurological risk by relaxing the neural structures and distraction must never be applied. This is a complex and potentially hazardous surgical salvage procedure which should never be necessary. All of these patients would have been much better treated by prophylactic surgery, to balance the growth of the spine, at a much earlier stage.

CONGENITAL KYPHOSIS AND KYPHOSCOLIOSIS

The classification of congenital kyphosis and kyphoscoliosis is very similar to that for congenital scoliosis. The kyphotic deformity may be due to either an anterior failure of formation of a vertebra or an anterior failure of segmentation of two or more vertebrae; there can be a mixture of these anomalies. In contrast to a congenital scoliosis, however, a kyphotic deformity, due to a failure of vertebral formation resulting in a posterior hemivertebra or a posterolateral quadrant vertebra,

progresses much more rapidly than one due to a failure of vertebral segmentation.

Congenital kyphosis and kyphoscoliosis are much less common than a congenital scoliosis but potentially more serious because they can, on occasion, lead to spinal cord compression and paraplegia. The reason for this is that a posterior hemivertebra or a posterolateral quadrant hemivertebra are unstable. As a result, the hemivertebra tends to extrude backwards into the spinal canal causing anterior spinal cord compression at the apex of the angular kyphosis, as the deformity increases.^{5,9} In contrast, neurological complications only occur in congenital scoliosis as a consequence of an associated intraspinal anomaly and are not caused by direct pressure on the spinal cord by the deformity.

Neurological complications and severe deformity can be prevented if the patient is treated prophylactically at an early age, when the kyphosis is small, by a posterior spine fusion to balance the growth of the spine. Delayed treatment for severe deformity or spinal cord compression requires much more difficult and potentially hazardous anterior and posterior spine surgery.^{6,9} The spine is approached anteriorly through a thoracotomy or thoraco-abdominal exposure to release the deformity and, if necessary, decompress the spinal cord combined with an anterior rib or fibula strut graft to obtain correction. Early stability may be obtained by a vascularised anterior rib strut graft. This is followed by a second stage posterior spine fusion with instrumentation to maintain correction.

Laminectomy and pre-operative traction are both absolutely contra-indicated if there is spinal cord compression. Traction initially corrects the mobile compensatory curves above and below the more rigid kyphosis. This lengthens the vertebral column and pulls the spinal cord against the unyielding apex of the kyphosis, resulting in further neurological deterioration. Laminectomy is ineffective because the spinal cord is not compressed posteriorly, and removal of the posterior bony structure destabilises the spine, resulting in more rapid progression of the kyphosis and greater anterior spinal cord compression.

SUMMARY

Congenital anomalies of the vertebrae producing a scoliosis, kyphoscoliosis or kyphosis, are potentially serious conditions, which can on occasion, result in an extremely severe rigid spinal deformity with malalignment of the body and possible spinal cord compression. Ideally, these anomalies should be diagnosed at an early stage when the curve is small. Congenital vertebral anomalies are frequently diagnosed in infants on radiographs taken for other reasons, before there is an obvious clinical deformity. These radiographic findings should not be ignored because they may provide an opportunity for early prophylactic treatment, before a clinical deformity develops. At this time, it is possible to anticipate prognosis based on the type and site of the vertebral anomalies, the degree of growth imbalance they produce and the amount of spinal growth remaining. Deterioration of the deformity must always be prevented. A deformity which is at risk of progression or spinal cord compression requires immediate prophylactic surgical treatment, no matter how young the patient. It is much better to carry out a

relatively simple operation to balance the growth of the spine at an early stage than to wait and perform potentially hazardous anterior and posterior spinal surgery, as a salvage procedure, when the deformity is severe or there is spinal cord compression.

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THE PEDIATRIC SPINE PRINCIPLES AND PRACTICE

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CONGENITAL SCOLIOSIS

MICHAEL J. MCMASTER

Congenital scoliosis is a lateral curvature of the spine caused by developmental vertebral anomalies that produce a lateral longitudinal imbalance in the growth of the spine. The term *congenital scoliosis* is slightly misleading because it implies that the curvature is present at birth, but this is not necessarily so. The vertebral anomalies are present at birth, but the clinical deformity may not become evident until later childhood, when scoliosis develops and the diagnosis is made radiographically. This type of scoliosis must not be confused with *infantile idiopathic scoliosis*, which can also present as a deformity in early childhood. In these patients, however, the spinal radiograph shows that there are no vertebral anomalies.

Congenital scoliosis is relatively uncommon, but the true incidence in the general population remains unknown because some vertebral anomalies produce so little deformity that they remain undetected.

The radiographic appearance of vertebral anomalies varies considerably. As a result, congenital scoliosis was for many years thought to be unpredictable in its behavior, and some thought that it seldom required treatment (2,6). In 1952, Kuhns and Hormell (35) reviewed 165 children with congenital scoliosis and concluded that this was usually a relatively benign condition that progressed slowly. It was not until 1968 that Winter and associates (68), in a major study involving 234 children, firmly established the much more serious prognosis for certain types of congenital scoliosis and described a radiographic classification based on the types of vertebral anomalies that cause it.

Ohtsuka and I studied the natural history of 251 patients with congenital scoliosis and found that all degrees of severity of curvature were seen at all ages (44). Some patients presented in their late teens with small curves that did not deteriorate significantly, whereas others presented in the first few years of life with curves that deteriorated rapidly, becoming severe deformities before the child reached the age of 10 years. These severe curves were often rigid, and surgical correction was difficult and dangerous. It is therefore important to be able to anticipate when a congenital scoliosis is at risk for rapid deterioration so that treatment can be initiated when the curve is small, rather than having to attempt dangerous surgical salvage procedures after the curve has become severe. Planning, such as prophylactic treatment, requires a thorough knowledge of the pathogenesis

and natural history of all types of congenital scoliosis and the various methods of treatment that are available.

ETIOLOGY

A family history of congenital scoliosis is unusual. Wynne-Davies (73) found that an isolated single anomaly, such as a hemivertebra, was usually sporadic, and there was no hereditary risk. Multiple vertebral anomalies and spina bifida cystica were etiologically related, however, and there was a 5% to 10% risk of either of these anomalies occurring in siblings or subsequent children. In contrast, Winter (64), found no significant relationship between multiple vertebral anomalies and relatives with spina bifida cystica. In addition, he found that an isolated single vertebral anomaly had about a 1 in 100 chance of occurring in a first-degree relative.

There are two rare hereditary conditions associated with congenital scoliosis in which most of the vertebrae in the thoracic and lumbar regions are malformed (55). In spondylothoracic dysostosis (Jarcho-Levin syndrome) (30), there are, in addition to the vertebral anomalies, also multiple posterior rib fusions that constrict the thorax and result in death from respiratory failure in early infancy. In spondylocostal dysostosis, the ribs are not so severely affected, and there is a normal life expectancy but with marked stunting of the trunk because of the vertebral anomalies.

Reports of congenital scoliosis in monozygotic twins have shown that if one twin has a vertebral anomaly, the other twin usually has a normal spine or less frequently an anomaly at a different level (23,51,52). Monozygotic twins have the same genetic material, and any difference in their vertebral development is likely to be due to an environmental insult rather than genetic factors. Mice embryos subjected to hypobaric hypoxia or carbon monoxide at an age equivalent to a 6-week-old human embryo also develop vertebral anomalies similar to those seen in congenital scoliosis (38,54). It can therefore be postulated that other environmental factors may have a similar effect on the human embryo vertebral column at this stage of its development.

PATHOGENESIS

To understand the natural history of congenital scoliosis and the great disparity in prognosis, it is necessary to correlate the

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principles of normal growth of the spine with the pathologic anatomy of the various types of congenital vertebral anomalies that may cause scoliosis. The embryologic development of the vertebral column is a complex and rapid process. The complete anatomic pattern is formed in mesenchyma during the first 6 weeks of intrauterine life (see Chapter 1). Defects of formation or segmentation of the primitive vertebrae occur during this period. After the abnormal mesenchymal mold is established, the cartilaginous and bony stages follow that pattern, and the vertebral anomalies are fully established at birth.

Normal longitudinal growth of the spine is the sum total of growth occurring at the end plates on the upper and lower surfaces of the vertebral bodies (5). This growth occurs symmetrically; as a result, the spine grows in a balanced manner without a pathologic curvature. In the presence of a congenital vertebral anomaly, however, there is an asymmetric deficiency in either the number of growth plates or their rate of growth, resulting in a longitudinal growth imbalance and an increasing spinal deformity as the child grows.

Congenital scoliosis, kyphoscoliosis, and kyphosis are part of a gradually blending spectrum of spinal deformities due to developmental anomalies that produce a localized imbalance in the longitudinal growth of the spine. The type of deformity that develops depends on whether the impaired growth occurs unilaterally producing a pure scoliosis or lies anterior or anterolateral to the transverse axis of vertebral rotation in the sagittal plane, producing kyphosis or kyphoscoliosis. Lordosis, caused by impaired posterior growth of the spine, is uncommon by itself and is usually associated with scoliosis. In a study of 584 consecutive patients with closed congenital spine deformities, I found that 472 patients had pure congenital scoliosis, 76 had kyphoscoliosis, and 36 had a pure kyphosis (48). Care was taken to distinguish between scoliosis with marked rotation, which could mimic kyphosis in the sagittal plane, and true kyphoscoliosis. This chapter deals only with those patients whose major deformity is scoliosis.

The rate of deterioration and final severity of congenital scoliosis are proportional to the degree of growth imbalance produced by the vertebral anomalies. This deterioration continues until skeletal maturity, when the growth plates fuse. However, the rate of spinal growth is not uniform. There are two periods of accelerated growth during which the scoliosis deteriorates more rapidly. The first occurs during the first 2 years of life, and the second occurs later during the adolescent growth spurt, which usually occurs in girls between the ages of 10 and 13 years and in boys about 2 years later. It is during these two periods that congenital scoliosis is most frequently diagnosed. On average, 25% of curves are nonprogressive, 25% progress slowly, and 50% progress more rapidly.

The neural axis and the vertebral column develop at the same time. It is therefore not surprising that neural and vertebral malformations often coexist and are termed *spinal dysraphism*. Abnormalities of the spinal cord are often associated with a failure of development of the posterior vertebral arches and overlying soft tissues, resulting in an "open" spinal defect, such as a myelomeningocele. This type of malformation often presents with major neurologic deficits in the lower limbs and paralysis of the trunk muscles, which contribute to the development of scoliosis.

This chapter deals only with "closed" congenital scoliosis in which the skin overlying the spine remains intact, although there may be mild neurologic abnormalities in the lower limbs as a result of occult intraspinal anomalies.

CLASSIFICATION AND NATURAL HISTORY

The major advances in the management of congenital scoliosis have been not in devising new methods of correcting severe deformities but rather in achieving a better understanding of the natural history of the condition and knowing when to apply prophylactic surgical treatment to balance the abnormal growth of the spine before there is a significant deformity.

The classification of the developmental vertebral anomalies that produce congenital scoliosis is based on the embryologic development of the spine. There are two basic groups of anomalies: those caused by a defect of formation and those caused by a defect of segmentation of one or more vertebrae (Table 1). About 80% of vertebral anomalies can be classified in this manner (Fig. 1). The remaining 20% are unclassifiable because there is a jumble of anomalies or the anomaly cannot be accurately identified, or because the scoliosis is so severe that it obscures the radiographic characteristics of the anomaly.

Vertebral anomalies may also be present in other areas of the spine outside the scoliotic curve, but these are ignored for the purposes of classification of the scoliosis if they do not contribute to the development of the deformity. Anomalies of the ribs are also common in association with congenital scoliosis, but these are not included in the classification because they do not cause scoliosis by themselves.

Defects of Vertebral Segmentation

Defects of vertebral segmentation may be either unilateral or bilateral and occur most frequently in the thoracic or thoracolumbar region.

Unilateral Failure of Segmentation

A unilateral failure of segmentation of two or more vertebrae results in a unilateral unsegmented bar, which is one of the most

TABLE 1. TYPES OF DEVELOPMENTAL VERTEBRAL ANOMALIES CAUSING CONGENITAL SCOLIOSIS

Defects of segmentation
Unilateral
Unsegmented bar
Unsegmented bar with contralateral hemivertebrae
Bilateral
Block vertebra
Defects of formation
Complete unilateral
Hemivertebra: fully segmented, semisegmented, nonsegmented, incarcerated
Partial unilateral
Wedge vertebra
Mixed or unclassifiable anomalies

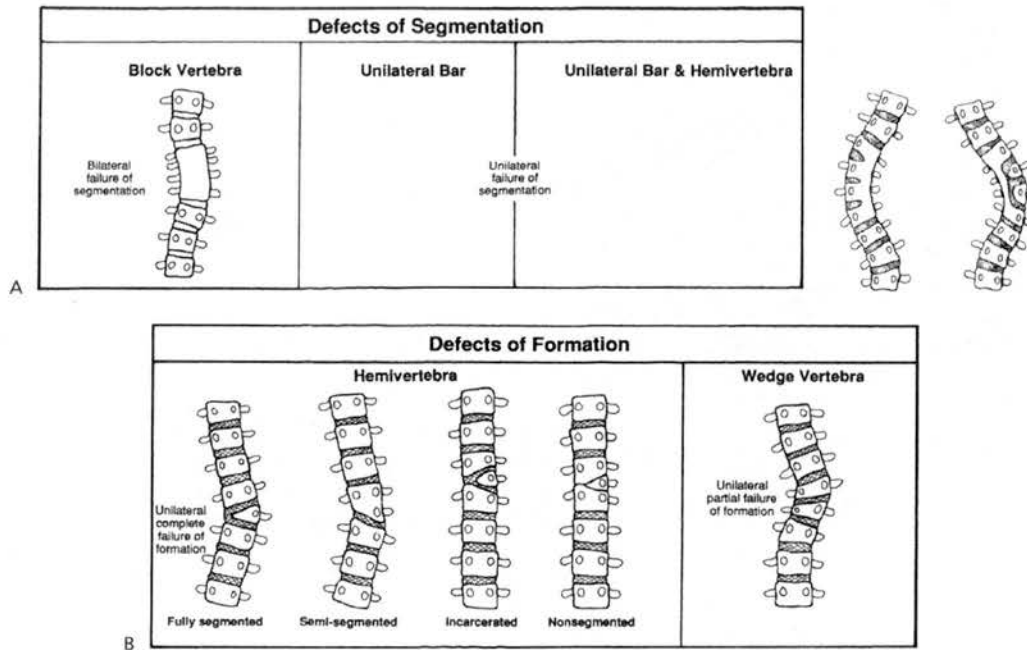


FIGURE 1. Congenital scoliosis.

common causes of congenital scoliosis (8,41,44,68). This type of anomaly consists of a bar of bone fusing the disc spaces and facet joints on one side of the spine while leaving the other side relatively unaffected. The unsegmented bar does not contain growth plates and therefore does not grow longitudinally. Some degree of growth continues on the opposite side of the spine, resulting in the development of scoliosis (Fig. 2). Rib fusions and other abnormalities of the ribs are often seen adjacent to the unsegmented bar, but they do not by themselves contribute to the development of the scoliosis. The rate of deterioration and the final severity of the scoliosis depend not only on the extent of the unsegmented bar but also on the growth potential on the convexity of the spine opposite the unsegmented bar. If the disc spaces on the convexity are well formed, it is more likely that the growth will be relatively normal, resulting in the development of a severe curvature. However, if the disc spaces are narrow and indistinct, there is likely to be impaired growth, resulting in a lesser curvature. On average, these curves deteriorate at a rate of 5 degrees a year, and most exceed 50 degrees by 10 years of age, resulting in a severe deformity.

There is also a small but important subgroup of patients who, in addition to the unsegmented bar, have one or more hemivertebrae on the convexity of the curve at the same level as the bar. Radiographically, this type of anomaly is seen most clearly in the first few years of life, but as the curve deteriorates and rotates, the hemivertebrae often become obscured by the deformity (Fig. 3). It is most important to recognize a unilateral unsegmented bar with contralateral hemivertebrae at an early

stage because this type of anomaly produces the most severe and rapidly progressive of all types of congenital scoliosis (47,50). Although there is no longitudinal growth on the side of the bar, the hemivertebrae produce a greater growth potential on the convexity than in patients with an unsegmented bar alone. On

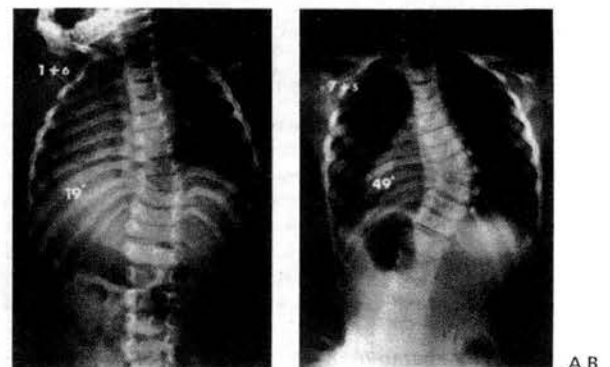


FIGURE 2. A: A boy aged 1 year, 6 months with a unilateral unsegmented bar on the left extending from T8 to T10 and producing a right thoracic scoliosis measuring 19 degrees. The disc spaces on the convexity opposite the bar are clearly seen, indicating a relatively normal growth potential; no growth is occurring in the unsegmented bar. No treatment was given. B: By the age of 7 years, 5 months, the thoracic curve had deteriorated to 49 degrees.

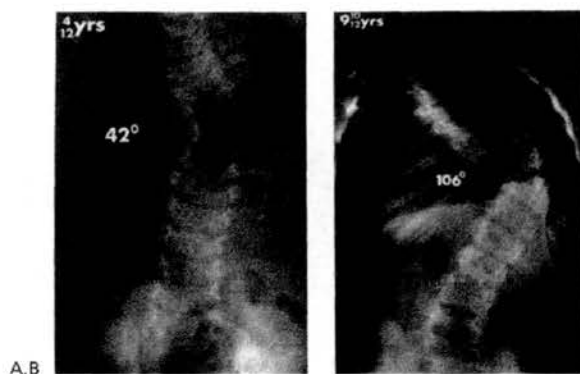


FIGURE 3. A: A 4-month-old girl with a 42-degree right thoracic scoliosis due to a unilateral unsegmented bar on the right extending from T7 to T10, with three adjacent contralateral hemivertebrae at the eighth to the tenth thoracic levels. No treatment was given. B: By the age of 9 years, 10 months, the scoliosis had increased to 106 degrees, and the contralateral hemivertebrae were no longer visible. There was marked malalignment of the trunk and distortion of the rib cage, reducing the vital capacity.

average, these curves deteriorate 6 degrees or more a year, and most exceed 60 degrees by 4 years of age. In addition, there is usually severe vertebral rotation and distortion of the rib cage due to the crankshaft effect produced by continuing growth of the hemivertebrae anterolaterally combined with the tethering effect of the unsegmented bar on the concavity of the curve. If untreated, these children become extremely deformed at an early age, and there is often significant respiratory impairment, which can lead to cor pulmonale.

In treating patients with an unsegmented bar with or without contralateral hemivertebrae, it is important to remember that it is not possible to create growth on the concave side of the spine where none exists. The only way to stop deterioration of the deformity is to prevent further growth on the convex side of the curve opposite the unsegmented bar, and this can only be accomplished by a spinal fusion.

Bilateral Failure of Segmentation

A bilateral failure of segmentation of a number of adjacent vertebrae results in a block vertebra. The disc spaces between the affected vertebrae are very narrow or fused. As a result, longitudinal growth is impaired on both sides of the spine, producing a shortened segment. When this occurs in the neck, it is part of the Klippel-Feil syndrome (25,70) and is usually diagnosed because of the short neck, low hairline, and restricted movement. A block vertebra occurring elsewhere in the spine is diagnosed less frequently because it produces little if any deformity. Occasionally, the longitudinal growth impairment is not symmetric, which can result in the development of lesser scoliosis that rarely exceeds 20 degrees and never requires treatment. Most commonly, the block vertebra is diagnosed as part of a complex of other congenital vertebral anomalies that may produce a more severe deformity.

Defects of Vertebral Formation

A lateral defect of vertebral formation can vary from mild wedging to the complete absence of half of the vertebra (i.e., a hemivertebra). These anomalies may affect one or more vertebrae and can occur in any part of the spine.

Unilateral Complete Failure of Formation

A hemivertebra is one of the most common causes of congenital scoliosis. It is caused by the complete failure of a vertebra to form on one side, resulting in a laterally based wedge of bone consisting of half a vertebral body, a single pedicle, and hemilamina. If the hemivertebra is in the thoracic region, there is usually an attached rib, resulting in an unequal number of ribs.

There are four different types of hemivertebrae, depending on the pathologic anatomy and relationship to the adjacent vertebrae (Fig 1). The hemivertebra may be fully segmented (nonin-carcerated), which is most common; semisegmented, which is less common; nonsegmented; or incarcerated, which is the least common. It is important to distinguish among these different types. Each has a different growth potential, and the severity of the resultant scoliosis is related to the degree of segmentation (46).

Fully Segmented Hemivertebra

A fully segmented (nonin-carcerated) hemivertebra has a normal disc space above and below and is completely separate from its adjacent vertebrae. Longitudinal growth occurs on the upper and lower surfaces of the hemivertebra, whereas there is an absence of two growth plates on the unformed side. As the hemivertebra grows, it acts as an enlarging wedge, producing scoliosis that usually deteriorates relatively slowly at a rate of 1 to 2 degrees a year.

A single fully segmented hemivertebra lies at the apex of the scoliosis, and as the curve deteriorates, the body of the hemivertebra tends to protrude slightly from the lateral margin of the spine. There may also be slight secondary wedging of the adjacent normal vertebrae (Fig. 4A, B). Lower thoracic and thoracolumbar curves deteriorate most rapidly and can exceed 45 degrees at skeletal maturity. However, there is often only a moderate cosmetic deformity because the hemivertebra does not have the same tethering effect on the concavity of the curve as a unilateral unsegmented bar and there is much less vertebral rotation (Fig. 4C).

A fully segmented hemivertebra at the lumbosacral junction is a particularly pernicious anomaly because it causes the lumbar spine to take off obliquely from the sacrum. This results in the development of long compensatory scoliosis in the thoracolumbar region. The secondary thoracolumbar curve, which does not contain any congenital anomalies, is initially mobile, but it later becomes fixed and rotated, with the upper body listing to the side opposite the hemivertebra (Fig. 5). This can produce a major cosmetic deformity and is best treated by prophylactic surgery at an early stage before the compensatory curve becomes fixed.

Two fully segmented hemivertebrae on the same side of the spine are less common but cause a much greater growth imbal-

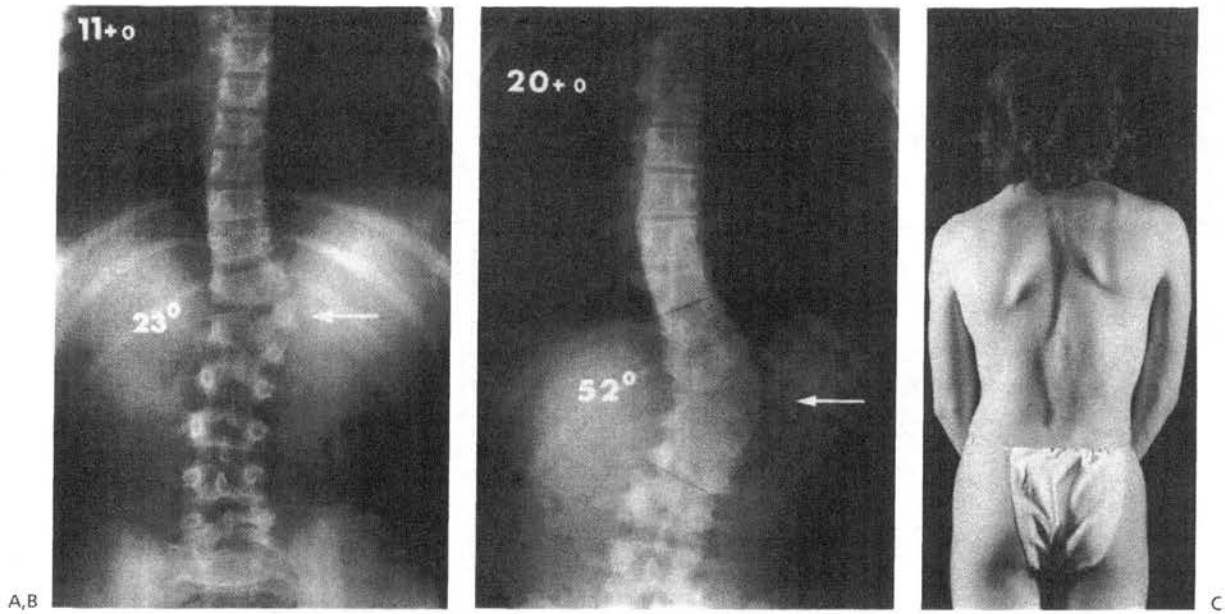


FIGURE 4. A: A boy aged 11 years with a 23-degree right thoracolumbar scoliosis due to a fully segmented (nonincurated) hemivertebra at L1. No treatment was given. B: By the age of 20 years, the patient was skeletally mature, and the curve had deteriorated to 52 degrees. C: Despite the severity of the curve, the cosmetic deformity was relatively mild.

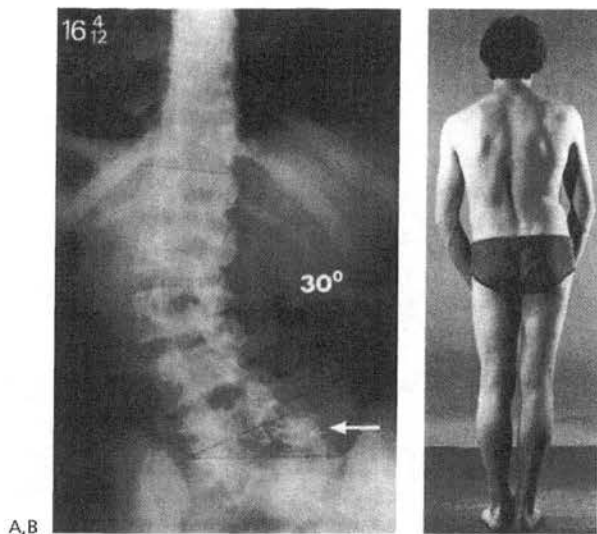


FIGURE 5. A: A boy aged 16 years, 4 months with a single unsegmented (nonincurated) hemivertebra at the lumbosacral junction, causing the lumbar spine to take off obliquely from the sacrum. A long secondary lumbar curve has developed, which measures 30 degrees and does not contain any congenital anomalies. B: The main cosmetic deformity is caused by the secondary lumbar curve and the tendency of the upper body to list to the left.

ance because there is an absence of four growth plates on the concavity of the curve. These curves deteriorate much more rapidly than those caused by a single hemivertebra. They deteriorate by about 3 degrees a year, and all exceed 50 degrees by 10 years of age (Fig. 6A, B). Without treatment, these curves could reach 70 degrees by skeletal maturity. Therefore, early prophylactic surgical treatment should be given as soon as they are diagnosed.

Two opposing fully segmented hemivertebrae are thought to be caused by an embryologic hemimetameric shift. The severity of the deformity depends on whether the hemivertebrae are close together or in different regions of the spine. If they are close together, separated by only one or two normal vertebrae, they tend to balance each other, causing only two small kinks in the spine. This produces minimal cosmetic deformity, and no treatment is required. If the hemivertebrae are in different regions of the spine, they produce separate curves that may be unbalanced, causing spinal decompensation and a significant cosmetic deformity (Fig. 7).

Semisegmented Hemivertebra

A semisegmented hemivertebra is synostosed to its neighboring vertebra and has only one disc space either above or below. As a result, two growth plates are obliterated on the convexity of the curve, which tends to balance the absence of the two growth plates on the unformed side of the hemivertebra. Although growth of the spine is theoretically balanced, the hemivertebra causes a tilting of the spine, which can induce slowly progressive scoliosis (Fig. 8). These curves usually do not exceed 40 degrees

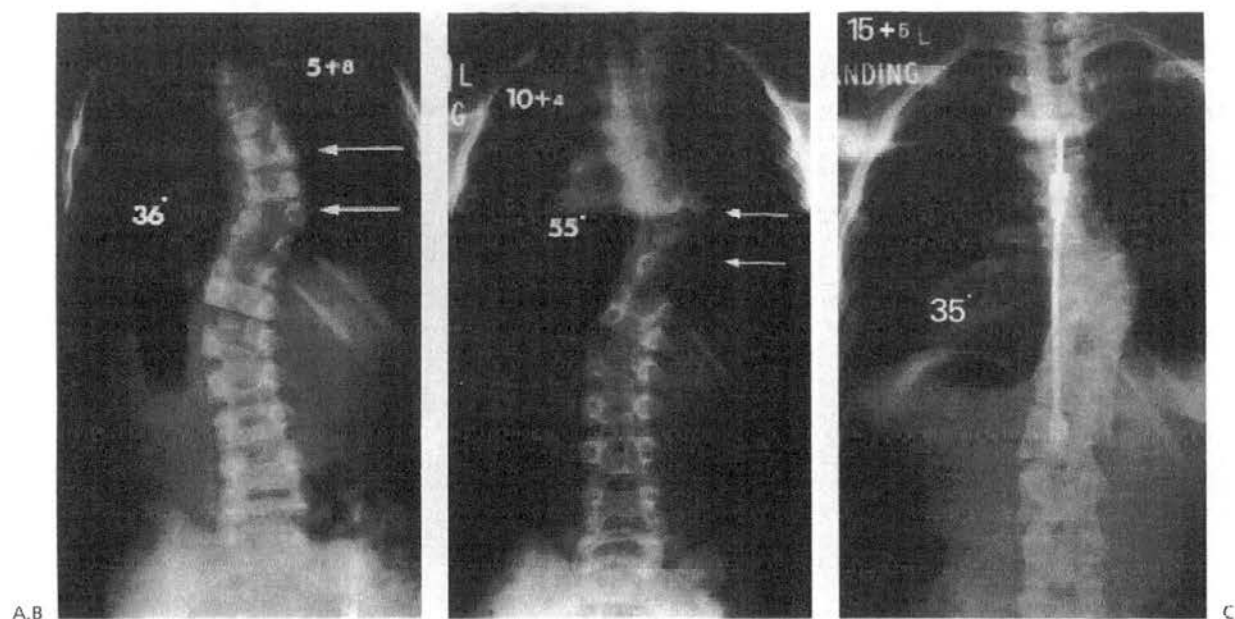


FIGURE 6. A: A girl aged 5 years, 8 months with a 36-degree right thoracic scoliosis due to two unilateral fully segmented (nonincurated) hemivertebrae at T7 and T9. No treatment was given. B: By the age of 10 years, 4 months, the thoracic curve had deteriorated to 55 degrees. At this time, the patient was treated by means of a posterior spinal fusion with instrumentation, which corrected the curve to 35 degrees. C: At 15 years, 5 months, there had been no deterioration.

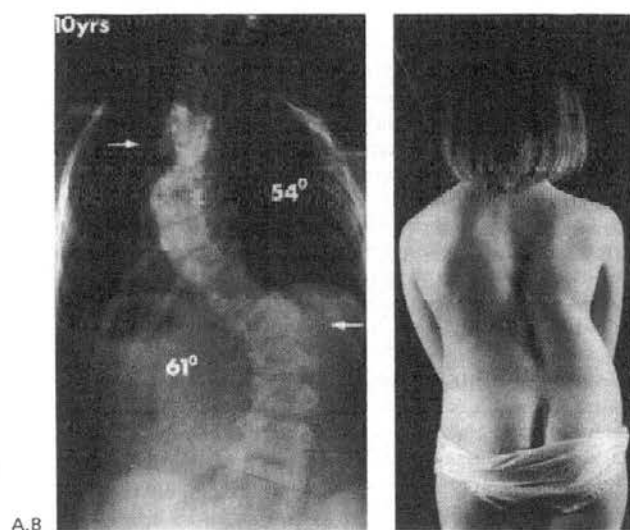


FIGURE 7. A: A girl aged 10 years with two opposing hemivertebrae occurring in different regions of the spine at T5 and L2. These hemivertebrae have produced two curves that are unbalanced. B: The patient lists to the left, and there is a significant cosmetic deformity.

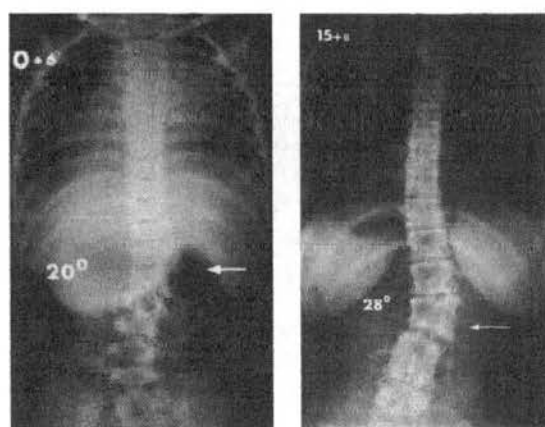


FIGURE 8. A: A girl aged 6 months with a 20-degree right lumbar scoliosis due to a semisegmented hemivertebra at L2. The hemivertebra is synostosed with L3. No treatment was given. B: The curve deteriorated very slowly to measure 28 degrees at 15 years, 8 months, when the patient was skeletally mature.

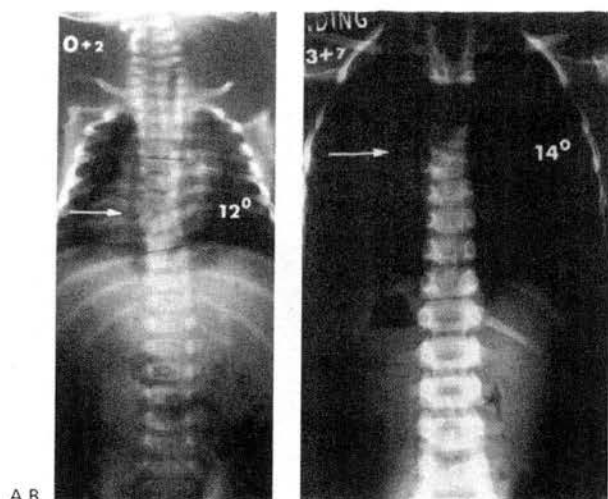


FIGURE 9. A: An infant aged 2 months with a minimal 12-degree left thoracic scoliosis due to an incarcerated hemivertebra at T6. No treatment was given. B: There was virtually no deterioration at 13 years, 7 months.

at skeletal maturity. Treatment is not usually required, except possibly when the semisegmented hemivertebra occurs at the lumbosacral junction.

Nonsegmented Hemivertebra

A nonsegmented hemivertebra is synostosed to both of its adjacent vertebrae and has no disc spaces either above or below. This type of hemivertebra, which usually occurs in the thoracic region, has no growth potential and therefore does not cause progressive scoliosis. There is no cosmetic deformity, and no treatment is required.

Incarcerated Hemivertebra

An incarcerated hemivertebra is more ovoid in shape and smaller than a fully segmented (nonincarcerated) hemivertebra. This type of hemivertebra is tucked into the side of the spine and set into a niche scalloped out of the adjacent vertebrae. The vertebrae above and below are shaped in such a way that they tend to compensate for the hemivertebra. As a result, the alignment of the pedicles on that side of the spine remains straight, and there is minimal scoliosis (Fig. 9). The disc spaces above and below the incarcerated hemivertebra are usually narrow and poorly formed, indicating a poor growth potential. The resulting scoliosis deteriorates very slowly if at all. Occasionally, the disc spaces become even narrower with growth and may fuse with the adjacent vertebrae. An incarcerated hemivertebra rarely produces scoliosis exceeding 20 degrees at skeletal maturity and never requires treatment.

Unilateral Partial Failure of Formation

A wedge vertebra is an uncommon cause of congenital scoliosis and is due to partial failure of a vertebra to form on one side.

The malformed vertebra contains two pedicles and is slightly wedged toward one side of the spine. There is retarded longitudinal growth on the hypoplastic side, which causes scoliosis that deteriorates relatively slowly. Surgical treatment may occasionally be necessary, especially if there are more than one wedged hemivertebra situated close together on the same side of the spine.

ASSOCIATED DEFORMING FEATURES

Apart from the severity of the congenital scoliosis, there are also a number of important secondary deforming features related to the site of the vertebral anomalies. These features contribute significantly to the overall disability and deformity of the patient and must be taken into consideration when planning treatment.

Upper thoracic curves are relatively common in congenital scoliosis. Although most never become very large, they can cause a significant cosmetic deformity because of elevation of the shoulder line and, less frequently, tilting of the head (Fig. 10). The higher the apex of the curve, the more severe the deformity. A 30-degree curve is probably at the upper limit of acceptability, especially in girls.

Midthoracic curves with their apex at T5, T6, or T7, especially those caused by a unilateral unsegmented bar with or without contralateral hemivertebrae, are frequently associated with the development of a long secondary structural curve in the lower thoracic or thoracolumbar region (Fig. 11). As the congenital scoliosis deteriorates, it produces a rotational effect that is transmitted further down the spine into the lower thoracic and thoracolumbar region, resulting in secondary scoliosis on the opposite side. This curve, which does not contain any congenital anomalies, is initially compensatory and correctable. Later, it becomes fixed and may deteriorate even more rapidly than the primary

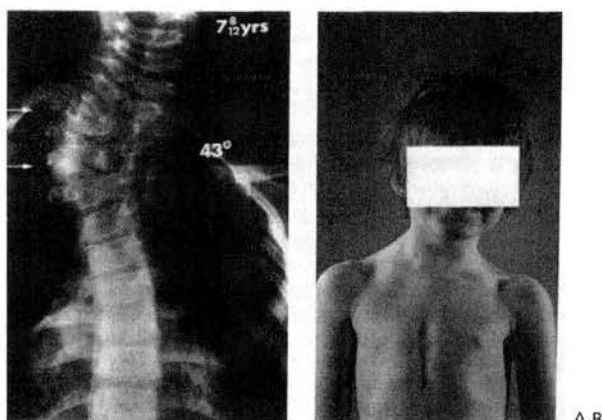


FIGURE 10. A: A girl aged 7 years, 8 months with a 43-degree right upper thoracic scoliosis due to two unilateral fully segmented (nonincarcerated) hemivertebrae at T2 and T4. B: Her main cosmetic deformity is the elevation of the right shoulder and slight tilting of the head to the left.

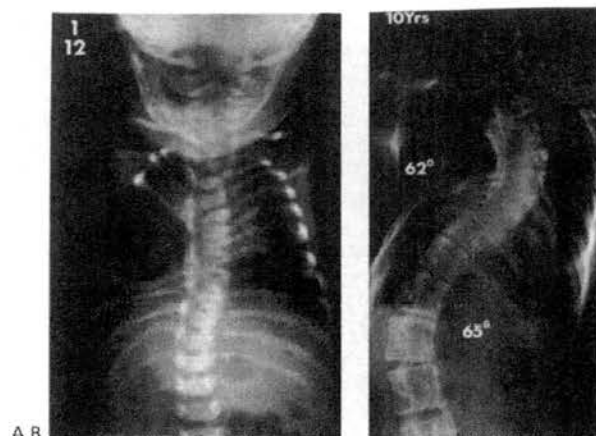


FIGURE 11. A: An infant aged 1 month who had a radiograph because of asymmetry of the thoracic cage. This showed that the spine was relatively straight, but there were absent ribs and a unilateral unsegmented bar extending from T3 to T7 on the left. No treatment was given. B: By the age of 10 years, the unilateral unsegmented bar had produced a 62-degree right upper thoracic scoliosis. However, the main deformity was caused by the secondary development of a severe left structural thoracolumbar scoliosis, which contained no congenital anomalies.

congenital curve. The congenital thoracic curve is often only moderately rotated, but the lower secondary curve is frequently severely rotated, producing the major cosmetic deformity of a large rib hump. In these circumstances, a severe, apparently "idiopathic" curve can develop below a congenital anomaly whose significance may not be fully appreciated.

Lower thoracic, thoracolumbar, and lumbar curves, especially those caused by a long unilateral unsegmented bar with or without contralateral hemivertebrae, often fail to develop compensatory curves either above or below that are sufficient to balance the congenital curve (Fig. 3). There are too few normal mobile vertebrae between the rigid anomalous segment and either the upper end of the spine or the sacrum. This results in a severe cosmetic deformity because, in addition to the spinal curvature, there is severe malalignment of the body, often associated with pelvic obliquity and an apparent leg length discrepancy.

Severe distortion of the rib cage frequently occurs secondary to the development of significant thoracic scoliosis. When this occurs before the child is 8 years of age, it interferes with the normal development of the lungs (13). Increasing deformity is associated with a steady decrease in vital capacity and may lead to cor pulmonale and possibly death in early adult life. Surgical treatment of the spine is associated with a further reduction in vital capacity, especially in patients with multiple thoracic anomalies (14).

ASSOCIATED OCCULT INTRASPINAL ANOMALIES

The development of the spinal cord is closely associated with that of the vertebral column. It is therefore not surprising that

neural and vertebral malformations often coexist. The spinal deformity may be obvious, but the intraspinal anomaly may not be recognized because the skin overlying the spine is intact and any associated neural deficit in the lower limbs may be mild or absent. It is important to detect these intraspinal anomalies because they may restrict movement of the spinal cord within the spinal canal and attempts to correct the spinal deformity could result in stretching of the neural tissues and serious complications.

A diastematomyelia is found in 5% to 21% of patients with congenital scoliosis (7,18,27,45,65). This is a sagittal split in a localized segment of either the spinal cord or cauda equina. Lying within the split is an osseous or fibrocartilaginous spur that projects backward from one or more adjacent vertebral bodies. The term *diastematomyelia* refers to the split in the neural structures and not to the spur or septum. It should not be confused with diplomyelia or true division of the spinal cord. Other types of congenital neural abnormalities—epidermoid cysts, dermoid cysts, neurenteric cysts, lipomas, and teratomas—may be found either alone or in association with diastematomyelia. The spinal cord or cauda equina may also be tethered by fibrous bands, a tight filum terminale, ectopic posterior nerve roots, or arachnoid adhesions to the dura or vertebral column.

In the past, myelography was used to diagnose these intraspinal anomalies. However, magnetic resonance imaging (MRI) is a much better method because it is noninvasive and more sensitive in the visualization of soft tissue abnormalities of the spinal cord. Using MRI, Bradford and coworkers (11) found a 38% prevalence of intraspinal anomalies in 42 patients with congenital spinal deformities; of these, 16 had a tethered cord, 4 had a diastematomyelia, 3 had a diplomyelia, 3 had a low-lying conus, 1 had a teratoma of the sacrum, and, surprisingly, 4 had an unsuspected syringomyelia. Winter and colleagues (72) found a similar prevalence of 41% in 48 patients.

Intraspinal abnormalities may be found in association with all sites and types of congenital scoliosis. However, the most common association (50%) is with a unilateral unsegmented bar with contralateral hemivertebra, producing scoliosis in the lower thoracic or thoracolumbar region (11,45).

An intraspinal anomaly may not be immediately obvious, but there are a number of clues to its presence. An abnormality of the skin overlying the spine, such as a dimple, nevus, hairy patch, or lipoma, is present in about 70% of affected patients. These stigmata, however, may also occur without an accompanying intraspinal anomaly (45). Neurologic abnormalities affecting the lower limbs may also be present, but they are often very mild and easily missed. Frequently, only one leg is affected. The leg may be slightly short with a small foot that may have a mild cavus deformity and slight clawing of the toes (29). In addition, the spinal radiograph may show a spina bifida occulta affecting one or more adjacent vertebrae associated with widening of the interpedicular distance and narrowing of the disc spaces. Occasionally, the bony spur associated with a diastematomyelia may be visible on the plain spinal radiograph, but an MRI is necessary to confirm its presence and reveal other possible neural abnormalities.

It is widely accepted that an intraspinal anomaly that tethers the spinal cord should be surgically released if there is a progres-

sive neurologic deficit or before attempting to correct a spinal deformity (49). Excision or release of the intraspinal anomaly will not improve the neurologic status, but it will prevent further deterioration and decrease the risk for complications after correction of the scoliosis. MRI is not necessary for all patients with congenital scoliosis, but it should be used in those with suspicious clinical or radiologic findings and in those who are about to undergo surgery to correct the spinal deformity. If the intraspinal anomaly is diagnosed during the routine assessment of children younger than 5 years of age, it should probably be removed as a prophylactic measure to prevent the possible development of neurologic deterioration. In older children, however, the presence of an intraspinal anomaly is not necessarily an indication for its removal unless there are symptoms or corrective surgery is planned.

OTHER CONGENITAL ANOMALIES

Congenital scoliosis is frequently associated with congenital anomalies in other systems, especially those formed from mesenchyma. These anomalies are often asymptomatic and may remain undetected until the patient is fully assessed after diagnosis of a congenital scoliosis. Beals and associates (3) found that up to 60% of patients with vertebral anomalies have one or more associated anomalies in other systems and that many of these are medically important. The prognosis for these patients is excellent if the associated anomalies are detected and, if necessary, treated.

The genitourinary system may be affected in up to 25% of patients. The most common anomalies diagnosed by means of an intravenous pyelogram (IVP) or ultrasound scan are a unilateral kidney, duplication of the kidney, or ureteric obstruction (15,43). The Klippel-Feil syndrome may be found in 25% (25,70) and congenital heart disease in 10% of patients (53).

Sprengel deformity (congenital elevation of the scapula) is often found in association with congenital scoliosis in the upper

thoracic or cervicothoracic region. When it is present on the convexity of the curve, the combination of these two anomalies causes a significant deformity because of elevation of the shoulder line. In these circumstances, the elevated scapula should also be reduced when the scoliosis is treated surgically. If the elevated scapula is on the concavity of the curve, however, it may partially compensate for the scoliotic deformity by leveling the shoulders. In these circumstances, it should not be reduced.

Goldenhar syndrome (oculoauricular vertebral dysplasia) is uncommon (19,20). Its main features are unilateral malformation of the ear and facial hypoplasia, and it is associated with congenital scoliosis, usually in the upper thoracic region.

PROGNOSIS

The prognosis for congenital scoliosis with regard to its rate of deterioration and final severity depends on three factors (Table 2):

1. *The type of vertebral anomaly and the degree of growth imbalance it produces.* The type of anomaly that causes the most severe scoliosis is a unilateral unsegmented bar with contralateral hemivertebrae at the same level. Next in severity is a scoliosis caused by a unilateral unsegmented bar alone, followed by two unilateral fully segmented hemivertebrae, a single fully segmented hemivertebra, and a wedge vertebra. The least severe deformity is caused by a block vertebra. Congenital scoliosis caused by a jumble of unclassifiable anomalies can be difficult to predict and requires careful monitoring. The poor prognosis associated with a unilateral unsegmented bar with or without contralateral hemivertebrae is so predictable that these curves should be treated immediately without any period of observation.
2. *The site of the anomaly.* For any type of vertebral anomaly, the rate of deterioration of the resulting scoliosis are most severe in the thoracic and thoracolumbar regions and are usually less severe in the upper thoracic and lumbar regions.

TABLE 2. MEDIAN YEARLY RATE OF DETERIORATION (IN DEGREES) WITHOUT TREATMENT FOR EACH TYPE OF SINGLE CONGENITAL SCOLIOSIS IN EACH REGION OF THE SPINE

Site of Curvature	Type of Congenital Anomaly					
	Block Vertebra	Wedge Vertebra	Hemivertebra		Unilateral Unsegmented Bar	Unilateral Unsegmented Bar and Contralateral Hemivertebrae
			Single	Double		
Upper thoracic	<1°-1°	★-2°	1°-2°	2°-2.5°	2°-4°	5°-6°
Lower thoracic	<1°-1°	2°-2°	2°-2.5°	2°-3°	5°-6.5°	5°-8°
Thoracolumbar	<1°-1°	1.5°-2°	2°-3.5°	5°-★	6°-9°	7°-14°
Lumbar	<1°-★	<1°-★	<1°-1°	★	>5°-★	★
Lumbosacral	★	★	<1°-1.5°	★	★	★

□ No treatment required □ May require spinal surgery □ Requires spinal fusion ★ Too few or no curves

Ranges represent the degree of deterioration before and after 10 years of age.

Modified from McMaster MJ, Ohtsuka K (1982): The natural history of congenital scoliosis: a study of 251 patients. *J Bone Joint Surg Am* 66: 588-601 and McMaster MJ (1998): Congenital scoliosis caused by a unilateral failure of vertebrae segmentation with contralateral hemivertebrae. *Spine* 23: 998-1005.

The site of the congenital scoliosis is also an important factor in the degree of cosmetic deformity produced, which can be tilting of the head, elevation of the shoulder line, decompensation of the trunk, and pelvic obliquity.

3. *The age of the patient at the time of diagnosis.* Congenital curves generally stabilize at skeletal maturity. A scoliosis presenting as a clinical deformity in the first few years of life has a particularly bad prognosis because this indicates a marked growth imbalance that will continue throughout the period of growth, resulting in a severe deformity. In addition, the rate of deterioration is not constant; it becomes much more severe after the age of 10 years, during the adolescent growth spurt. Even after skeletal maturity, severe curves may continue to deteriorate slowly as a result of plastic deformation of the spine.

TREATMENT

The objective of treatment in congenital scoliosis is to produce a spine that will be as straight as possible at the end of growth. It is not possible, however, to create growth on the concavity of the scoliosis, where it is either retarded or nonexistent. For patients with a marked spinal growth imbalance, there is no perfect treatment. The best that can be achieved is to balance spinal growth by retarding the growth on the convexity. In these circumstances, the optimal result is a short, relatively straight spine rather than the severely crooked spine that would have developed without treatment. There are three key factors in achieving an optimal result in patients with congenital scoliosis:

1. *Early diagnosis.* If the diagnosis is made early, while the curvature is still small, there is an opportunity for prophylactic surgery to balance the growth of the spine. Congenital vertebral anomalies are frequently diagnosed in infants on radiographs taken for other reasons, before there is an obvious clinical deformity (Fig. 11A). These radiographic findings should not be ignored. They may provide an opportunity for early prophylactic treatment before a deformity develops (Fig. 11B).
2. *Anticipation.* The prognosis for deterioration of congenital scoliosis can be anticipated based on the amount of spinal growth remaining, the type and site of the vertebral anomaly, and the degree of growth imbalance it produces. This requires careful study of good-quality spinal radiographs and a thorough knowledge of the natural history of the condition. Often, the most helpful radiographs are those taken at an early stage before the vertebral anomalies are obscured by the developing deformity (Fig. 3).
3. *Prevention of deterioration.* It is easier to prevent a severe deformity than to correct one. Some types of congenital scoliosis, such as those caused by a unilateral unsegmented bar with or without contralateral hemivertebrae, have such a bad prognosis that no observation is necessary; they require immediate surgical treatment no matter how young the patient. Other types of congenital scoliosis may be observed, but one of the most common errors is to fail to recognize slow and relentless progression until it is too late for prophylactic treat-

ment. All patients require careful radiologic assessment at 4- to 6-month intervals. After progression is established, immediate treatment is necessary to prevent further deterioration. A simple operation to balance the growth of the spine and prevent increasing curvature is preferable to a hazardous multistage surgical procedure to correct a severe and rigid deformity at a later stage.

Radiographic Assessment

Spinal radiographs are necessary to identify the type of vertebral anomaly that is causing the congenital scoliosis and to monitor any deterioration of the deformity.

Identification of the vertebral anomalies requires good-quality supine anteroposterior and lateral spinal radiographs centered on the abnormal levels. The diagnosis of a congenital scoliosis is made on the anteroposterior spine radiograph. It is also important to examine carefully the lateral radiograph to detect any associated kyphosis because this will affect the treatment. If only the anteroposterior radiograph is viewed, a patient with a posterolateral quadrant vertebra producing kyphoscoliosis may be misdiagnosed as having a lateral hemivertebra producing scoliosis. Treatment by means of a combined anterior and posterior convex epiphysiodesis, which may be appropriate for a lateral hemivertebra, would be contraindicated for kyphoscoliosis because it would increase the deformity in the sagittal plane. A misdiagnosis is also possible in infants with a short unilateral unsegmented bar, which may not appear radiographically until it is fully ossified. A coned-down oblique radiograph of the apex of the curve gives a true anteroposterior view of the more severely rotated spine and may detect hidden vertebral anomalies. An anteroposterior view aligned through the lumbosacral junction shows whether a lumbosacral hemivertebra is fully segmented or semisegmented. An MRI scan may also help define the type of vertebral anomaly.

After the vertebral anomalies have been clearly identified on the anteroposterior radiograph, it should be possible to count the number of vertebral growth plates on both sides of the spine and estimate the potential growth imbalance. Allowance should be made for any disc space that is narrowed or ill defined because this indicates an impaired growth potential. It may be difficult to assess the prognosis in young infants with mixed vertebral anomalies; they require careful radiologic follow-up to detect deterioration of the scoliosis.

Follow-up radiographic assessment of congenital scoliosis does not require detailed visualization of the vertebral anomalies after they have been fully identified. Serial full-length standing anteroposterior spinal radiographs are best for assessing deterioration. The Cobb angle of the scoliosis is always measured at exactly the same vertebral levels with reference to the standing radiograph taken when the patient was first seen (39). These radiographs show any deterioration of the scoliosis as well as any change in shoulder symmetry, decompensation of the trunk, or pelvic obliquity.

Nonoperative Treatment

Certain types of congenital scoliosis can occasionally be managed by the application of an orthosis, but this is never an alternative

to appropriate surgical treatment. The problem being treated is one of spinal growth imbalance, and an orthosis should never be applied if there is a severe growth imbalance or a rigid curve such as that produced by a unilateral failure of segmentation.

In young children with congenital scoliosis, the application of a plaster jacket or underarm brace in an attempt to prevent deterioration of the deformity constricts the chest and may have an adverse effect on the developing respiratory system. Winter and colleagues (67) found that the Milwaukee brace does not have the same adverse effects on pulmonary function. It has the advantage of being a nonconstricting device yet is able to apply corrective forces to selected areas of the trunk. This type of brace works best on long flexible thoracic or thoracolumbar curves with a short anomalous segment, such as a hemivertebra at the apex of the curve. A Milwaukee brace is unlikely to be effective if the scoliosis is greater than 40 degrees or if there is less than 50% flexibility assessed on side-bending or distraction radiographs.

If the brace does not control the congenital curve, it is pointless to continue its use. The patient requires immediate surgery. After surgery, a brace may be helpful in controlling spinal alignment and the development of compensatory curves that were not included in the fusion. These secondary structural curves are easier to control than primary congenital curves because they occur in areas of the spine where the vertebrae are relatively normal and therefore much more mobile than the anomalous segment, which is often rigid. Tilting of the head may be controlled in a Milwaukee brace with a head extension. An underarm brace may be sufficient for an adolescent with a secondary thoracolumbar or lumbar curve.

Surgical Treatment

Surgical treatment is frequently necessary for congenital scoliosis and is indicated at any age if the deformity is severe or the curve is increasing and cannot be controlled. There is no one operative procedure that can be applied to all types of deformities. The method of surgery selected depends on the age of the patient, the site and type of vertebral anomaly, and the size of the curvature. Successful surgical treatment depends on selecting the right procedure and applying it at the right time.

Prophylactic surgical procedures are applied to young patients presenting with small curves that have a bad prognosis. The objective of treatment is to prevent further deterioration by balancing the growth of the spine. After prophylactic surgery, these curves must be carefully followed to skeletal maturity because further surgical treatment may be required later.

Salvage surgical procedures are necessary for patients who present at a later stage with larger deformities that require correction. The primary objective is still to prevent further deterioration, and correction is attempted only if it can be safely achieved.

Prophylactic Convex Growth Arrest (Hemiepiphyodesis)

A convex growth arrest procedure (combined anterior and posterior convex hemiepiphysiodesis) is the optimum form of early prophylactic surgical treatment for patients with a small progres-

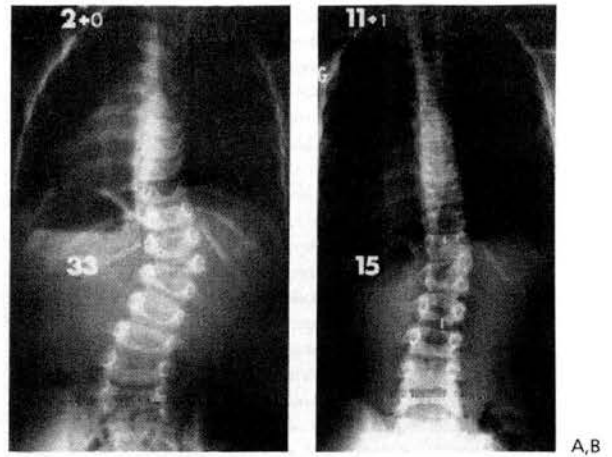


FIGURE 12. A: An infant aged 2 years with a 33-degree right thoracolumbar scoliosis due to a single fully segmented (nonincurved) hemivertebra at L1. A combined anterior and posterior convex growth arrest procedure was performed. B: By the age of 11 years, 1 month, the curve had improved to 15 degrees.

sive scoliosis due to a unilateral failure of vertebral formation in which there is some growth potential on the concavity at the site of the vertebral anomaly. This procedure is best applied to patients younger than 5 years of age with a short curve caused by a fully segmented hemivertebra that corrects to less than 40 degrees, with the patient supine (1,9,59,61,63,71) (Fig. 12). The objective of the surgery is to balance the growth of the spine by preventing further unbalanced growth on the convexity at the site of the hemivertebra. Theoretically, this should allow the scoliosis to correct slowly by means of the continuing growth on the concavity. This is a relatively safe procedure; the only disadvantage is the slow and often uncertain correction because of the unpredictable growth potential on the concavity of the curve. Even if there is no correction of the deformity, however, a convex growth arrest is often sufficient to stabilize the deformity.

This procedure does not result in correction of the scoliosis in patients with a unilateral failure of vertebral segmentation because there is no potential for growth on the concavity of the curve. The presence of any degree of kyphosis is also a contraindication to the procedure because the anterior convex growth arrest aggravates the deformity.

The surgery is performed in two stages that are usually carried out under the same anesthetic. The spine is first approached anteriorly on the convexity of the scoliosis through a thoracotomy, a thoracoabdominal retroperitoneal exposure, or a purely retroperitoneal approach beneath the diaphragm, depending on the site of the hemivertebra. The lateral half of the discs and their adjacent end plates are removed not only at the site of the hemivertebra but also at one intervertebral level above and below. This removes the anterior growth plates at the site of the anomaly, which are the main cause of the increasing scoliosis. To create an anterior convex fusion, the excised disc spaces are packed with chips of bone taken from an excised rib. The second

stage of the procedure is performed through a separate posterior exposure of the convexity of the curve at the site of the hemivertebra. Care is taken not to strip the paraspinal muscles on the concavity of the curve and possibly interfere with the growth potential on this side of the spine. A posterior convex fusion is performed by excising the facet joints and decorticating the posterior elements not only at the site of the hemivertebra but also to one intervertebral level above and below. Strips of excised rib are applied to the fusion area. Several days after surgery, partial correction may be attempted by means of manual traction and the application of a plaster jacket, which is maintained for 6 months until the convex growth arrest has soundly healed.

A transpedicular anterior convex epiphysiodesis, which can be performed at the same time as the posterior convex epiphysiodesis, has been described as an alternative that obviates the need for a separate anterior approach (31,34). The pedicle of the hemivertebra and the pedicles of the vertebrae above and below are entered on the convexity. Using a curette, the hemivertebra is decancellated along with the convex half of the vertebrae above and below. Curved curettes are used to perforate the end plates of the vertebrae to be included in the hemiepiphysiodesis. Strips of autogenous iliac crest bone graft are then placed through the pedicles and into the decancellated portion of the vertebral bodies and packed across the disc spaces. This technique has the potential advantage of reducing the morbidity associated with a separate thoracotomy or thoracoabdominal retroperitoneal approach. There are as yet no long-term follow-up reports of this procedure, however, and the results can only be fully assessed at skeletal maturity.

Minimally invasive thoracoscopic and laparoscopic procedures have recently gained considerable clinical interest and can also be used to perform an anterior convex epiphysiodesis or arthrodesis as well as excise the body of a hemivertebra. These procedures have the potential to reduce postoperative pain and recovery time, and they produce a more cosmetically pleasing result because of small incisional scars. There is a considerable learning curve associated with these techniques, however, and as yet, there are no long-term results.

Excision of a Hemivertebra

Excision of a fully segmented hemivertebra is theoretically attractive as a prophylactic procedure because it removes the primary cause of the scoliosis, which is the enlarging wedge on the convexity at the apex of the curve (12,56,62). This also creates a wedge osteotomy of the spine, which, when closed, produces correction.

The major advantage of resecting a hemivertebra is that it allows maximal correction of the deformity and realignment of the spine. Most single hemivertebrae, however, do not cause significant spinal imbalance when seen at an early stage. Most surgeons, therefore, use the simpler combined anterior and posterior convex growth arrest procedure, which is a less risky operation, but it has the relative disadvantage of less predictable correction that depends on continuing growth on the concavity of the curve. If there is a more severe deformity, excision of the hemivertebra and realignment of the spine can be a more beneficial, although more hazardous, procedure.

The hemivertebra is excised in two stages. The spine is approached both anteriorly and posteriorly in a manner similar to that for a convex growth arrest procedure. In the first stage, the body of the hemivertebra and the anterior part of the pedicle are removed along with the adjacent discs and vertebral end plates, and the dura is visualized. In the second stage of the procedure, the spine is exposed posteriorly, and the posterior elements of the hemivertebra are removed, along with the transverse process and posterior part of the pedicle. This is a technically demanding procedure with a risk for direct injury to the spinal cord and bleeding from the epidural veins (26). It has also been suggested that the dissection necessary to resect the hemivertebra may interfere with the segmental blood supply to the spinal cord (32). To prevent possible ischemic neurologic complications, some surgeons have advocated that the two stages be separated by 10 days to allow time for the vasculature to recover (33,36,64). However, it has now been established that successful results can be achieved when both procedures are performed under the same anesthetic (4,10).

After the hemivertebra has been excised, the scoliosis can be corrected by closing the wedge osteotomy. During this procedure, the spine remains relatively stable because the ligaments have been maintained on the concavity and act as a stabilizing hinge as the wedge is closed. However, correction may be difficult to obtain in the thoracic region because of the supporting rib cage. Correction is easier and safer in the more mobile lumbar spine, where the cauda equina is also more resilient. Spinal compression instrumentation applied to the convexity of the curve above and below the excised hemivertebra may be used posteriorly to close the wedge osteotomy (4,26). This requires strong bone to seat the hooks. Care must be taken not to compress the emerging nerve root as the osteotomy is closed, especially at the lumbosacral junction. It may be difficult or dangerous to apply this type of instrumentation to very small children. In these situations, correction may be easier and safer to obtain by the postoperative application of a double-pantaloon spica cast; this may be wedged at a later stage to obtain further correction (10). Regardless of the method of correction, the patient must wear a spica cast for 3 months after the surgery. This should be followed by 3 months in a brace until the osteotomy is soundly healed.

The objective of excising the hemivertebra is to achieve a balanced spine and prevent curve progression. The operation should be performed early while the scoliosis is still relatively mobile and before the development of secondary structural changes in the curve. The best long-term results after resection of a hemivertebra are achieved if the scoliosis can be nearly completely corrected. It is usually possible to obtain up to 30 degrees of correction of lumbar curves. Larger curves, especially those that have already developed secondary structural changes or fixed compensatory curves, do not fully correct and often continue to deteriorate. This may be partially prevented by extending the surgery to include an anterior and posterior convex growth arrest of the disc spaces one level above and below the excised hemivertebra. Failure to fuse the whole length of the scoliosis often leads to a slow progressive loss of correction, which necessitates further posterior surgery to extend the fusion when the child is older.

In my opinion, the best indication for hemivertebra excision is its occurrence at the lumbosacral junction, where it causes an oblique take-off of the lumbar spine and a major spinal imbalance. The only way of realigning the trunk is by excising the hemivertebra and closing the osteotomy (57). This is successful only at a relatively early stage before the secondary thoracolumbar compensatory curve becomes fixed.

Prophylactic Early Spine Arthrodesis In Situ

Arthrodesis in situ is the optimal form of early prophylactic surgical treatment for a congenital scoliosis caused by a unilateral failure of vertebral segmentation when seen at an early stage before there is significant deformity. A convex growth arrest procedure would not correct this type of deformity because there is no growth potential in the unsegmented bar on the concavity of the curve. These anomalies produce very rigid deformities with a known potential for severe progression. After significant scoliosis has developed, it can be corrected only by a spinal osteotomy, which can be a difficult and hazardous procedure. The object of an early spine arthrodesis is to stabilize the curve at an early stage by creating a solid thick fusion that will stop the unbalanced growth of the spine. The arthrodesis should be performed as soon as the anomaly is recognized, and the best results are achieved when this is carried out before the age of 2 years (47,66). The argument that an early spine fusion will stunt the growth of the spine is of no relevance with this type of congenital scoliosis. The abnormal segment is not contributing to vertical height, and it is only making the spine more crooked. It is much better to achieve a short, relatively straight spine that is balanced than a spine that is even shorter because of the severe curvature.

The congenital scoliosis contains not only the unilateral unsegmented segment but also a number of relatively normal vertebrae at the upper and lower ends that are also tilted into the curve. Failure to include all of these vertebrae in the fusion results in continued progression of the deformity (47). With this type of congenital scoliosis, the surgeon is unlikely to regret operating too early or fusing too many vertebrae but will always regret delaying surgery or having fused too few vertebrae.

Loss of correction after an early posterior spinal fusion in infantile and juvenile idiopathic scoliosis has been attributed to either a weak fusion mass or increasing rotation of the spine resulting from the crankshaft effect produced by continuing unbalanced anterior growth of the spine in the presence of the posterior tether caused by the fusion (16,24). This phenomenon, however, does not usually occur to a great extent in congenital scoliosis because the anterior growth plates in the anomalous segment are also frequently abnormal. A posterior arthrodesis alone can produce an acceptable result in congenital scoliosis, provided it results in a strong, thick fusion capable of overcoming the anterior growth of the spine (58). The crankshaft effect is seen only in those patients who already have marked vertebral rotation as part of the deformity (40).

In my experience, it is usually advisable to perform a single-stage combined anterior and posterior spine arthrodesis if there is a severe growth imbalance, such as that produced by a unilateral unsegmented bar with contralateral hemivertebra (47). This pro-

cedure has the advantage of directly overcoming any possible crankshaft effect and producing a more solid stable fusion. The rib excised at the time of the thoracotomy provides a good source of autogenous bone graft, which may not be available from the iliac crest in these very young children. Postoperatively, a spinal jacket is applied for 6 months to allow the fusion to heal.

The secondary structural thoracolumbar scoliosis, which can develop below congenital thoracic scoliosis with its apex at T5, T6, or T7, also requires prophylactic treatment. This secondary deformity is not controlled by an early fusion of the primary congenital scoliosis. Bracing may control the secondary curve and postpone extending the fusion to the lower lumbar region, especially if many years of further growth are anticipated. Alternatively, it is possible at the time of the posterior arthrodesis of the congenital curve to apply pediatric spine instrumentation across both curves without fusing the secondary curve. Subsequent serial extension of the instrumentation allows additional longitudinal growth in the unfused thoracolumbar curve before extending the fusion when the child is older.

Correction and Posterior Spinal Arthrodesis

Attempted correction of the deformity and posterior spinal arthrodesis is the usual surgical procedure for an older child with a moderately severe congenital curve that is still relatively flexible (Fig. 13). The object of surgery is to achieve overall balance of the spine rather than excessive correction of the congenital curve. The posterior arthrodesis is performed over the whole length of the deformity from the upper to the lower neutral vertebrae and not just at the anomalous segment. Although it is not usually possible to obtain correction at the site of the anomalous vertebrae, moderate correction may be achieved at the relatively normal vertebral levels that lie above and below this area and are still within the scoliosis. Correction may be attempted either intraoperatively by the insertion of instrumentation at the time of the posterior spinal arthrodesis or postoperatively by the application of a spinal jacket.

An MRI scan is essential in all patients before intraoperative correction of the deformity is attempted. This may reveal the presence of an intraspinal anomaly, such as a diastematomyelia, that could be tethering the cord. If this is not removed, the patient could develop serious neurologic complications if a distraction force is applied to the spine to correct the deformity (Fig. 13C). Because of the complexity of the two operative procedures, it is best to have the intraspinal anomaly removed during a separate neurosurgical procedure performed several weeks before the attempted correction of the spinal deformity and posterior arthrodesis.

The safest method of correcting the deformity is to apply a spinal jacket 5 days after the posterior spinal arthrodesis. The patient is awake during this procedure, and the jacket is applied on a traction frame. With this technique, there is negligible risk for neurologic complications. The disadvantages are that it produces relatively little correction, and the patient has to wear a cumbersome spinal jacket extending from the neck to the pelvis for 6 months until the fusion is solid. This method is best applied to young children whose bones are too soft or underdeveloped to allow the effective application of internal fixation.

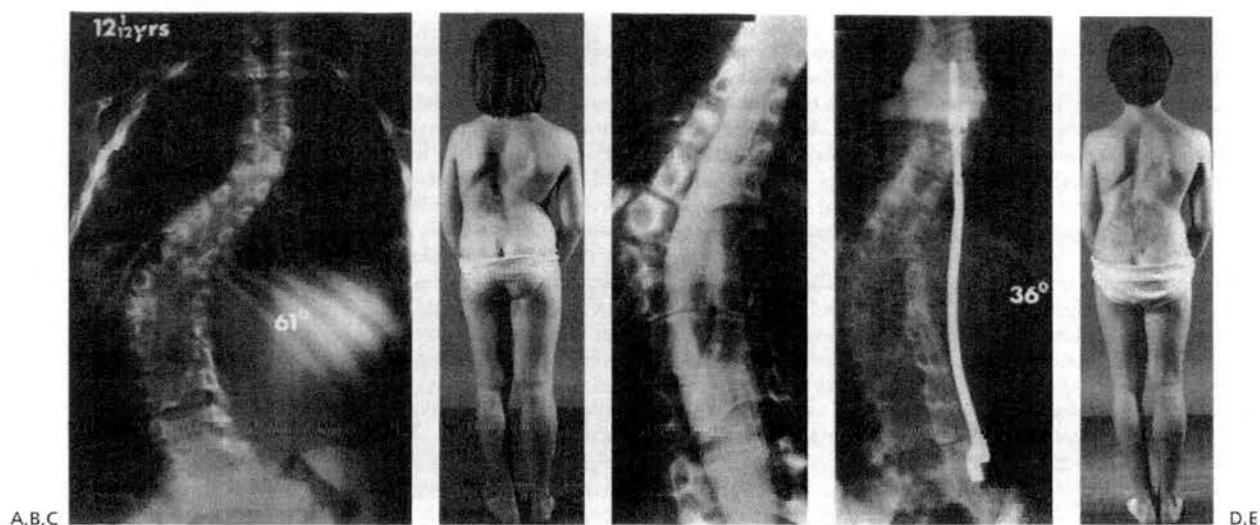


FIGURE 13. A: A girl aged 12 years, 1 month with a 61-degree thoracolumbar scoliosis due to complex multiple vertebral anomalies. The spinal radiograph also shows a spina bifida occulta affecting a number of adjacent vertebrae in the thoracolumbar region, associated with widening of the interpedicular distance and narrowing of the disc spaces. B: The patient had a hairy patch at the thoracolumbar junction, and there was slight shortening and hypoplasia of the left leg. C: A myelogram shows a diastematomyelia at the thoracolumbar junction. D: The diastematomyelia was excised, and the scoliosis was corrected to 36 degrees with Harrington instrumentation. E: At the age of 15 years, there is a solid fusion with no loss in correction and a good cosmetic result.

The use of posterior spinal instrumentation to correct a congenital scoliosis at the time of the spinal arthrodesis has several advantages: It achieves moderately better correction and reduces the incidence of pseudarthrosis when compared with a posterior spinal fusion in a spinal jacket alone (21,69). Spinal instrumentation does, however, carry a greater risk for neurologic complications because of the effect of distraction on the spinal cord while the patient is anesthetized. Of the various types of scoliosis, the congenital variety carries the highest risk for neurologic complications after intraoperative correction. Arthrodesis and instrumentation without correction, other than that achieved passively while the patient is lying on the operating table, should be used if there is any suspicion of an intraspinal anomaly, even if it has been removed before the surgery. The instrumentation should be used only as an internal strut to support the spine and aid the development of a solid fusion (42).

Despite the risks, spinal instrumentation is frequently used to obtain moderate correction in patients with congenital scoliosis, but specific measures must be taken during the surgery to detect the possible development of neurologic complications. Spinal cord monitoring using evoked potentials is essential, but unfortunately it is not completely reliable (17). The wake-up test (60) or the ankle clonus test (28) should be performed immediately after correction of the deformity. These tests, however, monitor only the current neurologic status, and it is possible for neurologic abnormalities to develop at a later stage in the operative procedure (22,37).

In the past, Harrington instrumentation has been used most

frequently in the treatment of congenital scoliosis (21,69). Newer forms of segmental spinal instrumentation using multiple hooks, pedicle screws, or sublaminar wires are more controversial in their application. Congenital abnormalities of the laminae may make it difficult to apply these types of instrumentation. It may also be dangerous to pass wires or hooks into the spinal canal at the site of the anomalous segment, which could be congenitally narrowed or contain an abnormal spinal cord. Regardless of the method of instrumentation, it is important to remember that the main objective is to achieve trunk balance rather than excessive correction of the scoliosis, which may be dangerous. If there is any suspicion that a neurologic deficit may have developed during the surgery, the spinal instrumentation should be removed immediately and the patient treated postoperatively in a plaster jacket.

Anterior and Posterior Spinal Osteotomy, Vertebrectomy, Correction, and Arthrodesis

A spinal osteotomy is a major salvage procedure that theoretically should never be necessary in congenital scoliosis. Its use is indicative of a failure to apply the basic principles of early diagnosis, anticipation, and prevention of increasing deformity. Patients who require a spinal osteotomy would have been much better treated at an earlier stage by simpler surgical procedures.

The indication for a spinal osteotomy is a severe rigid deformity with either fixed pelvic obliquity or marked decompensation of the upper trunk relative to the pelvis (Fig. 14). These

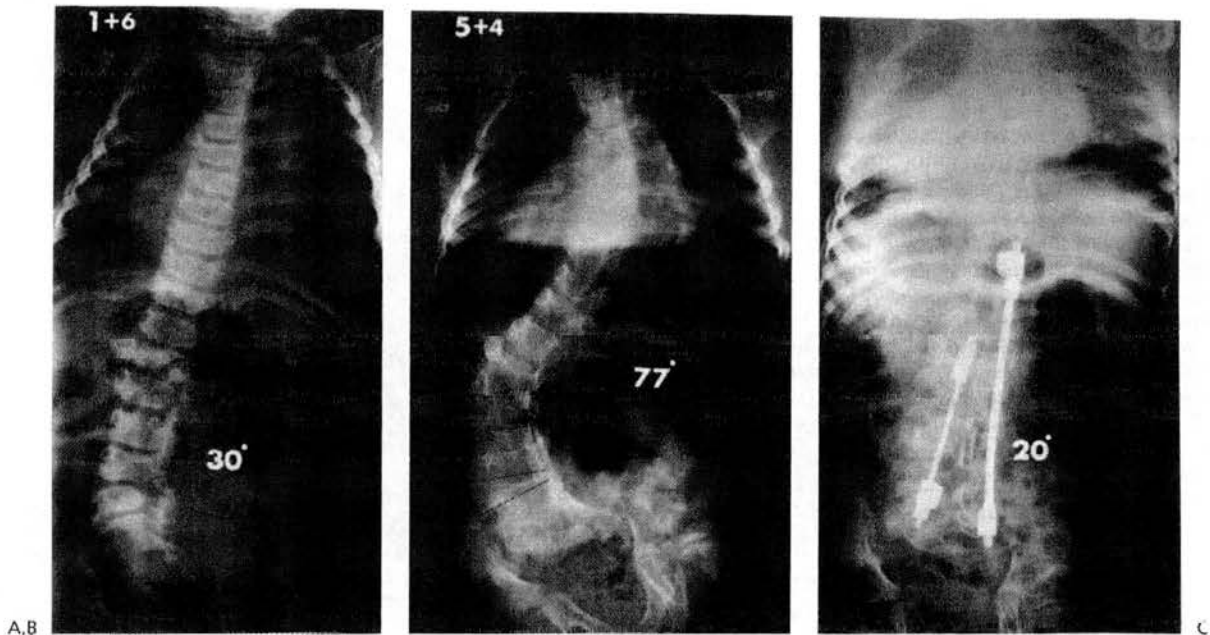


FIGURE 14. A: An infant girl who was radiographed at 1 year, 6 months and found to have 30-degree left lumbar scoliosis due to a unilateral unsegmented bar on the right extending from L4 to the sacrum, with a contralateral hemivertebra at L6. No treatment was given. B: By the age of 5 years, 4 months, the scoliosis had increased to 77 degrees, resulting in a severe rigid deformity with translocation of the trunk and marked pelvic obliquity. C: Two-stage anterior and posterior spinal osteotomies were performed at two levels in the lumbar region, and the scoliosis was corrected to 20 degrees with posterior spinal instrumentation.

severe spinal deformities are usually caused by a unilateral unsegmented bar with or without contralateral hemivertebrae.

The object of the surgery is to osteotomize the spine both anteriorly and posteriorly to create mobility, which will allow correction and enable the head and upper trunk to be balanced over a level pelvis. This is a technically demanding procedure with a significant neurologic risk and should be performed only by an experienced spinal surgeon after full consultation with the patient or parents and only if there are no alternatives.

The surgery can be performed as a single combined procedure or in two stages separated by 5 to 7 days. In the first stage, the spine is exposed anteriorly through a thoracotomy or a thoracoabdominal retroperitoneal approach, depending on the site of the vertebral anomalies. The intervertebral discs and their associated end plates are excised over the whole length of the deformity. The unsegmented bar on the concavity is then osteotomized through the sites of the excised discs. Simply osteotomizing the unsegmented bar will not allow the spine to grow straight because there is no growth potential on that side of the curve. To obtain significant correction, it may also be necessary to perform a wedge resection of a vertebral body at the apex of the curve, with the base of the wedge on the convexity and tapered toward the concavity. This is a difficult and dangerous procedure that is often associated with significant blood loss from the raw bony surfaces and epidural veins.

In the second stage of the procedure, the whole length of the congenital scoliosis is exposed posteriorly. The sites of the anterior osteotomies are identified, and posterior osteotomies are performed at the same levels by removing part of the lamina and facet joints on the convexity and dividing the unsegmented bar on the concavity. If an anterior vertebrectomy has been performed at the apex of the curve, it will be necessary to carry out a posterior wedge osteotomy at the same level by excising the lamina and removing the pedicle and transverse process on the convexity. The safest and best site for a vertebrectomy is in the lumbar region, where it is easier to close the wedge because there is no interference from the rib cage and the cauda equina is more resilient than the spinal cord.

After the anterior and posterior osteotomies have been performed, correction is obtained by applying posterior compression instrumentation to the convexity to close the wedge osteotomies. This shortens the spinal column and relaxes the neural structures, decreasing the risk for complications as well as straightening the scoliosis and balancing the spine (36). Only after this has been achieved is it safe to apply instrumentation to the concavity—not to achieve further correction but to act as a supporting strut and to maintain correction over the whole length of the deformity. A posterior spinal arthrodesis is performed over the entire length of the scoliosis extending from the upper to the lower neutral vertebrae and not just at the site

of the osteotomy. Postoperatively, a plaster jacket or spica is applied to help stabilize the spine until the fusion is solid.

SUMMARY

Congenital scoliosis is a potentially serious condition that can result in an extreme deformity with malalignment of the body. Ideally, this type of scoliosis should be diagnosed at an early stage when it is possible to anticipate the prognosis based on the amount of spinal growth remaining, the type and site of the vertebral anomalies, and the degree of growth imbalance they produce. Deterioration of the deformity must always be prevented. Scoliosis that is at risk for progression requires immediate prophylactic surgical treatment, no matter how young the patient. It is much better to carry out a relatively simple operation to balance the growth of the spine at an early stage than to wait and perform a dangerous surgical salvage procedure when the deformity is severe.

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Section C

Infantile Idiopathic Scoliosis

INFANTILE IDIOPATHIC SCOLIOSIS: CAN IT BE PREVENTED?

MICHAEL J. McMASTER

From the Edinburgh Scoliosis Unit

The changing incidence of idiopathic scoliosis in 672 patients who attended the Edinburgh Scoliosis Clinic between 1968 and 1982 inclusive is reported. Of these patients, 144 had infantile, 51 juvenile and 477 adolescent idiopathic scoliosis. Thirty-seven of the infantile curves progressed and 107 resolved. The relative frequency of both progressive and resolving infantile idiopathic scoliosis (given as a percentage of the total number of patients with idiopathic scoliosis) declined from 41.75 per cent in the four years from 1968 to 1971, to four per cent in the three years from 1980 to 1982. It is suggested that infantile idiopathic scoliosis is a preventable deformity and that the position in which the infant is laid may be a causative factor.

Idiopathic scoliosis is a structural deformity without any evident clinical cause. Depending on the age of presentation it has been classified into three types: infantile (presenting from birth to three years), juvenile (presenting from 3 to 10 years) and adolescent (presenting from 10 years to skeletal maturity). Wynne-Davies (1968) carried out a family survey of all types of idiopathic scoliosis seen in Edinburgh. She found that there was a genetic tendency to develop scoliosis but this by itself was insufficient: other factors (still unknown) were necessary to "trigger off" the deformity. Infantile idiopathic scoliosis differs from the adolescent variety in several important ways. Infantile scoliosis is more common in boys, and the great majority of patients have a left thoracic curve. During the juvenile years there is a gradual change in the clinical pattern until by adolescence the majority of patients are girls and a high proportion have a right thoracic scoliosis. The most striking difference, however, is that between 52 and 90 per cent of infantile curves resolve spontaneously (Lloyd-Roberts and Pilcher 1965; Ferreira and James 1972) whereas adolescent curves do not resolve and nearly all progress (Ponseti and Friedman 1950; James 1954). These findings suggest that although all idiopathic curves may have a common genetic basis, the unknown triggering factors in infancy may be very different from those in adolescence.

Surveys in North America have confirmed the genetic nature of idiopathic scoliosis (Cowell, Hall and MacEwen 1972) but have revealed major differences in the relative frequencies of the different types. The Edinburgh survey showed that 50 per cent of all patients with idiopathic scoliosis had infantile curves whereas in Boston, USA, only 0.5 per cent of idiopathic curves were

infantile (Riseborough and Wynne-Davies 1973). Since the white populations of Edinburgh and Boston stem from similar genetic stock, the different frequencies of infantile idiopathic scoliosis suggests that either some beneficial aspect of the environment of North America actively prevents its development or that some adverse triggering factor is present in Great Britain but absent in North America.

Since 1972 I have noted a marked change in the number of patients presenting to the Edinburgh Scoliosis Clinic with infantile idiopathic scoliosis. This clinic is the only referral point for all patients with scoliosis from a large area of Scotland and therefore reflects any change in the relative frequencies of the different types of idiopathic scoliosis in this population. The purpose of this paper is to report on this change and comment on its possible significance.

Table I. The number of girls and boys presenting with idiopathic scoliosis between 1968 and 1982

Type of idiopathic scoliosis	Number of patients		
	Male	Female	Total
Infantile	85	59	144
Juvenile	18	33	51
Adolescent	96	381	477
Total	199	473	672

MATERIALS AND METHODS

A retrospective study was made of all patients under the age of 18 years presenting for the first time with idiopathic scoliosis to the Edinburgh Scoliosis Clinic between January 1968 and December 1982. Only patients from our usual referral area in Scotland, for which there is no other referral point, were included. There were 672 patients. They were classified into three groups depending on the age of onset of their curve: 144 patients with

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infantile idiopathic scoliosis (65 of whom were seen before 1972 and have been previously investigated by Wynne-Davies, 1975); 51 patients with juvenile idiopathic scoliosis; and 477 patients with adolescent idiopathic scoliosis. All patients in the juvenile and adolescent groups had progressive curves of over 20 degrees. The sex ratio in each of the three groups is shown in Table I.

RESULTS

Incidence of scoliosis. The yearly incidence of patients presenting with infantile, juvenile and adolescent idiopathic scoliosis is shown in Figure 1.

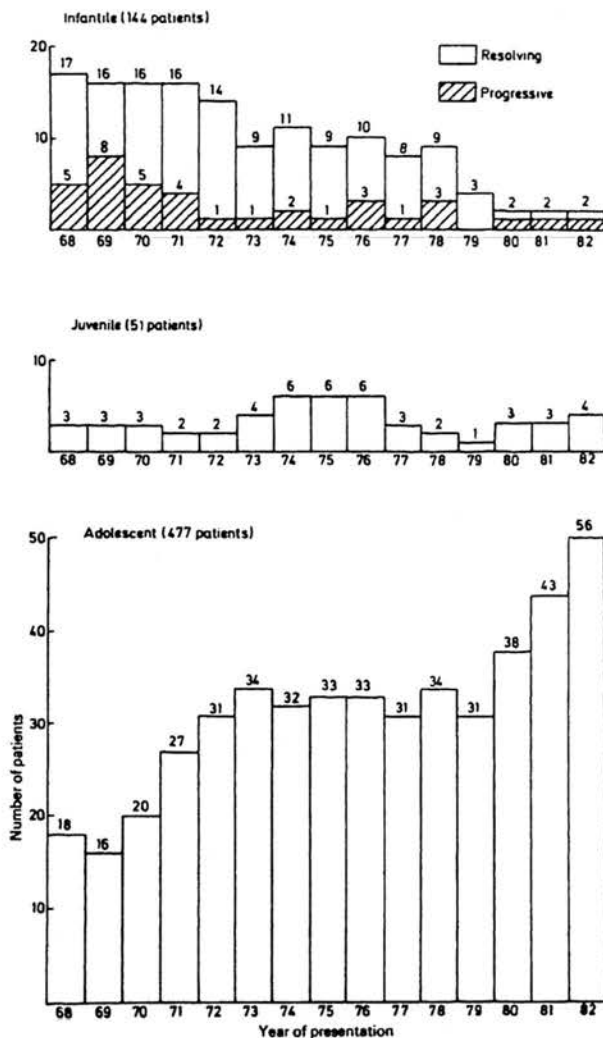


Fig. 1

The yearly incidence of patients presenting with infantile, juvenile and adolescent idiopathic scoliosis between 1968 and 1982.

Infantile. From 1968 to 1971 the number of patients presenting with infantile idiopathic scoliosis remained constant at 16 to 17 per year. Of the 65 patients diagnosed during these four years, 43 (66 per cent) had curves which

resolved and 22 (34 per cent) had curves which progressed. After 1971 there was a steady decline in the number of patients presenting with infantile scoliosis with only six patients presenting between 1980 and 1982. Of these six patients, three had resolving curves and three had progressive curves. The declining incidence of infantile idiopathic scoliosis has therefore applied to both progressive and resolving curves.

Juvenile. The yearly incidence of juvenile idiopathic scoliosis remained fairly constant with between one and four patients presenting each year from 1968 to 1982 (except from 1974 to 1976 when there was a slight increase with six patients presenting in each year).

Adolescent. The yearly incidence of adolescent idiopathic scoliosis increased from 18 patients in 1968 to 31 patients in 1972. Between 1972 and 1979 there was a fairly constant rate with 31 to 34 patients presenting each year. In the years 1980 to 1983 the yearly rate of incidence again increased with 38, 43 and 56 patients presenting respectively.

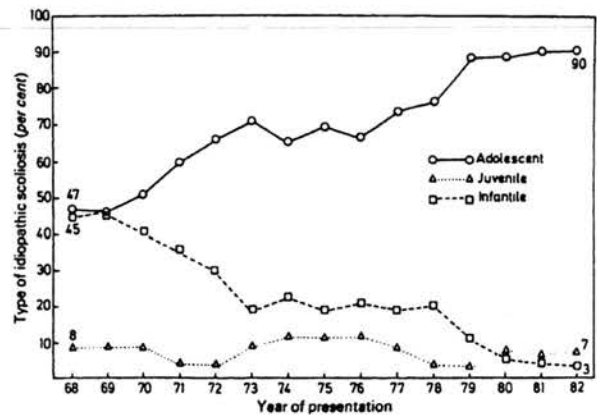


Fig. 2

Graph illustrating the relative frequency of infantile, juvenile and adolescent idiopathic scoliosis from 1968 to 1982.

The relative frequency of the different types of idiopathic scoliosis from 1968 to 1982 is shown in Figure 2. Of the patients presenting with idiopathic scoliosis in 1968, 47 per cent had adolescent curves and 45 per cent infantile curves. There was then a steady decline in the frequency of infantile curves and a corresponding increase in the frequency of adolescent curves until 1982 when 90 per cent of the patients with idiopathic scoliosis had adolescent curves and three per cent infantile curves. The frequency of juvenile curves between 1968 and 1982 remained fairly constant at around eight per cent.

Infantile idiopathic scoliosis. Of the 144 patients with infantile idiopathic scoliosis 107 (74 per cent) had curves which resolved spontaneously and 37 (26 per cent) had curves which progressed. The clinical details of these patients are shown in Table II. The ratio of boys to girls was 3:2 and was the same for both progressive and resolving curves.

One hundred and twenty-one infants (84 per cent) developed curves in the first six months of life; of these patients 83 per cent had curves that resolved and only 17 per cent had curves that progressed. In contrast, 30 per cent of the 23 children whose scoliosis was diagnosed after one year of age had curves that resolved and 70 per cent had curves that progressed.

A single structural scoliosis occurred in 138 patients (96 per cent); 130 infants had thoracic curves, seven had thoracolumbar curves and only one had a lumbar curve. Eighty-two per cent of these single curves were left-sided and only 18 per cent right-sided. Seventy-eight per cent of single curves resolved and 22 per cent progressed.

A double structural scoliosis affecting the thoracic and lumbar regions was less common and occurred in only six infants (four per cent). Double curves were most frequently diagnosed after the first year of life. They were equally distributed on either side of the spine and all progressed, but more slowly than single progressive curves.



Fig. 3

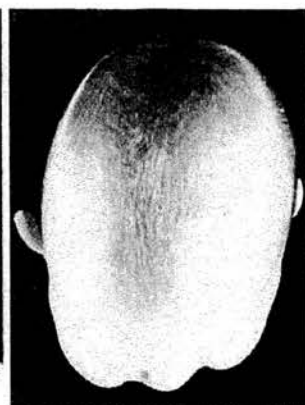


Fig. 4

Left-sided plagiocephaly with contralateral bat ear.

with the convex side of the curve in all but four patients.

All patients with progressive infantile idiopathic scoliosis required treatment (McMaster and Macnicol 1979).

DISCUSSION

Over the last 10 years there has been an increasing awareness of the need for early recognition and referral of all types of scoliosis to a specialised clinic where accurate diagnosis and treatment can be carried out. As a result, increasing numbers of patients with idiopathic scoliosis have presented to the Edinburgh Scoliosis Clinic but, paradoxically, there has been a marked decline in the number with infantile idiopathic scoliosis and this has applied to both progressive and resolving curves (Fig. 1). Progressive infantile curves are never overlooked because they produce a severe deformity at an early age, and it is very unlikely that more resolving curves are now being missed than in the past. A similar decline in the previously high incidence of infantile idiopathic scoliosis has been observed in West Germany (Mau 1981).

If we look at the relative frequency of the different types of idiopathic scoliosis presenting each year, we see an even more dramatic change (Fig. 2). Of the 157 patients with idiopathic scoliosis seen between 1968 and 1971; 41 per cent had infantile curves, seven per cent juvenile curves and 52 per cent adolescent curves. All the juvenile and adolescent curves progressed, whereas only 34 per cent of infantile curves progressed and 66 per cent resolved. Since 1971 there has been a continuous gradual change in the relative frequency of the different types of idiopathic scoliosis. The frequency of adolescent scoliosis has increased, the frequency of juvenile scoliosis has remained static and that of infantile scoliosis has decreased markedly. Of the 153 patients with idiopathic scoliosis seen between 1980 and 1982 only four per cent had infantile curves, seven per cent had juvenile curves and 90 per cent had adolescent curves. All the juvenile and adolescent curves progressed whereas only three of the infantile curves progressed and three resolved. The relative frequency of the different types of idiopathic

Table II. Clinical details of 144 patients with infantile idiopathic scoliosis

	Number of patients		Total	Percentage
	Progressive curves	Resolving curves		
<i>Site of curvature</i>				
Thoracic	28 (22L, 6R)	102 (85L, 17R)	130	90
Thoracolumbar	2 (1L, 1R)	5 (4L, 1R)	7	5
Lumbar	1 (L)	none	1	1
Double	6*	none	6	4
<i>Sex</i>				
Male	24	61	85	59
Female	13	46	59	41
<i>Age at presentation</i>				
Birth to 6 months	16	82	98	68
7 months to 1 year	5	18	23	16
1 year to 3 years	16	7	23	16
<i>Plagiocephaly</i>				
Present	26	98	124	86
Absent	2	7	9	6
Unknown	9	2	11	8

*3 right thoracic, left lumbar; and 3 left thoracic, right lumbar

Plagiocephaly (Figs 3 and 4) was present in 124 infants (86 per cent) and absent in nine (six per cent), all with resolving curves; no clinical record had been made in the remaining 11 infants (eight per cent). In the patients with progressive curves, either single or double, the "recessed" side of the plagiocephaly always corresponded with the convex side of the thoracic or thoracolumbar curve. In the patients with resolving curves, the "recessed" side of the head corresponded

scoliosis seen in Edinburgh now approaches that found in North America: a survey in Boston of 208 patients with idiopathic scoliosis found that 0.5 per cent had infantile curves (one patient), 10.5 per cent had juvenile curves and 89 per cent had adolescent curves (Riseborough and Wynne-Davies 1973). The frequency of adolescent idiopathic scoliosis in the two cities is now virtually identical, but infantile scoliosis is still more common in Edinburgh than in Boston and juvenile scoliosis is less common (Fig. 5).

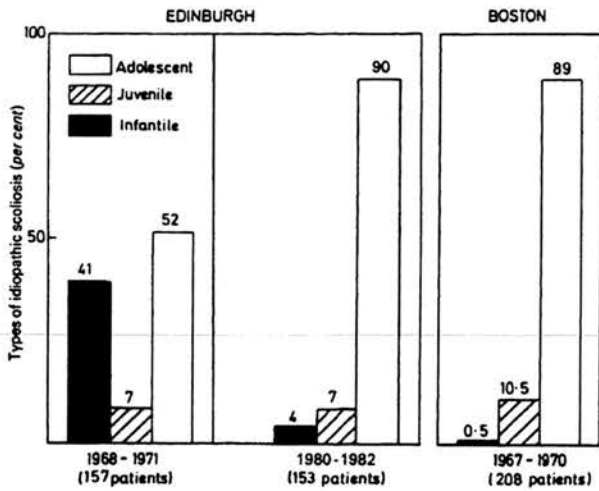


Fig. 5

Bar chart showing the incidence of idiopathic scoliosis in Edinburgh and Boston.

Wynne-Davies (1975) suggested that the aetiology of infantile idiopathic scoliosis is multifactorial, with a genetic tendency to develop the deformity which is "triggered off" by different medical or environmental factors. The balance in each infant is likely to be different, an infant with a strong genetic tendency needing very little triggering action and vice versa. From this study it would appear that although the genetic tendency to develop infantile idiopathic scoliosis remains constant, the triggering factors are being removed, or alternatively some beneficial factor presumably present in the environment of North America is now being applied in Edinburgh.

There is a very close association between plagiocephaly and both resolving and progressive infantile idiopathic scoliosis. Plagiocephaly is a plastic deformation of the skull. It is thought to occur when an immobile infant habitually lies towards one side. The action of gravity on the plastic skull causes the uppermost side of the face and head to flow backwards and become recessed; the lower ear is pushed forwards producing the commonly associated contralateral "bat ear" (Fig. 6). This deformity is not usually present at birth (Hay 1971) but was found in 28 per cent of 223 normal infants examined in Edinburgh between the ages of six weeks and six months. The recessed side of the head was on the

left in 83 per cent (Wynne-Davies 1975). The incidence of plagiocephaly in 63 premature Edinburgh babies was 63 per cent; this higher incidence is thought to be due to their greater immobility (Brown, Bell and Fulford 1982). Plagiocephaly, however, rarely persists and once the child becomes mobile it usually resolves by the age of six years. In my study of infantile idiopathic scoliosis, plagiocephaly occurred in 96 per cent of the children seen in the first six months of life and in 86 per cent of all the children. The recessed side of the head was on the left in 81 per cent. The scoliosis in these infants was rarely noted at birth but, like the plagiocephaly, developed within the first six months of life in 70 per cent. The convex side of the curve corresponded with the recessed side of the head in all except four infants with resolving curves. This close association between the presence, time of presentation and side of the deformity of both plagiocephaly and infantile idiopathic scoliosis suggests a common pathogenesis. Wynne-Davies (1975) found that in infants with idiopathic scoliosis there was a greater than normal incidence of prematurity, low birth-weight, mental retardation (in boys), older mothers, poorer homes, congenital heart disease, and delayed maturation of muscles and ligaments indicated by herniae and congenital dislocation of the hip (Hooper 1980). All of these factors contribute to immobility of the newborn and, when combined with posture, may trigger the development of scoliosis in a genetically predisposed infant.

Observations in Edinburgh (Brown *et al.* 1982) and elsewhere (Mau 1972) have shown that during the first few months of life there is a natural tendency for all infants to turn towards their right side when laid supine. This would account for the predominance of left-sided plagiocephaly in both normal infants and those with scoliosis. In my opinion, when an immobile infant is laid supine and partly turns towards the right side, the

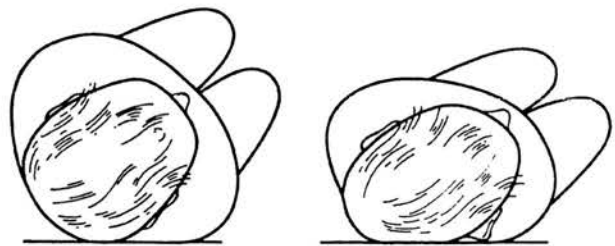


Fig. 6

Diagram illustrating postural moulding of the skull producing left-sided plagiocephaly and contralateral bat ear. Note the moulding of the thorax.

immature thorax also undergoes plastic deformation due to the influence of gravity. The uppermost hemithorax, like the skull, tends to flow backwards causing the thoracic vertebrae to rotate backwards on their long axis (Figs 7 and 8). In the normal spine there is a biomechanical coupling of axial rotation and lateral bending. White (1978) has stated that axial rotation of the midthoracic

vertebrae can on occasion induce a lateral curvature of the spine in which the anterior aspects of the vertebrae point towards the convexity of the curve. Backward vertebral rotation in the infant could therefore induce an upward curvature of the spine and a left-sided thoracic scoliosis in infants lying on their right side. The tendency to produce an upward curve is increased by pressure from the mattress on the ribs of the lower hemithorax. Once the central nervous system and the vertebral musculature mature, the infant becomes much more mobile and the

idiopathic scoliosis. The prone position is theoretically advantageous because the child cannot roll onto its side and this prevents asymmetric moulding of the thorax and promotes early maturation of the spinal extensor muscles. In the past, infants in Great Britain were usually laid on their back or towards one side and were often wrapped tightly in blankets. However, in the last six to eight years there has been an increasing tendency to advise mothers to nurse their babies prone and, because more people now have central heating, there is less need to wrap

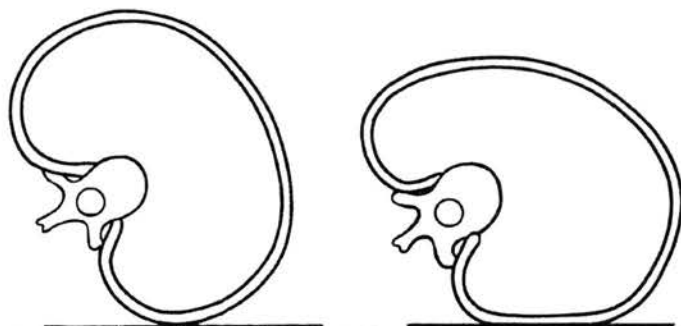


Fig. 7

Figure 7—Diagram illustrating postural moulding of the thorax when a baby is laid supine and partly turned towards its side. Figure 8—A baby boy aged four months who presented with a left thoracic idiopathic scoliosis. He has habitually been laid supine and partly turned towards his right side resulting in moulding of his head and trunk. The creases on his trunk indicate his relative immobility. The head shows a left-sided plagiocephaly and there is a right bat ear which has been against the mattress.

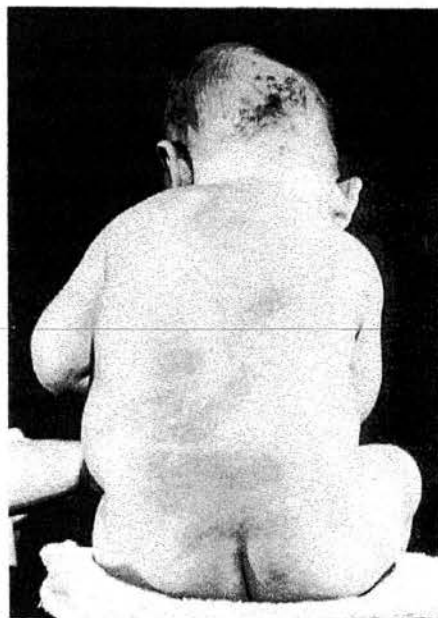


Fig. 8

tendency to spinal curvature is usually overcome; this accounts for the high incidence of resolving curves. If, however, there is a marked vertebral rotation associated with the genetic, medical and environmental factors discussed earlier, it is possible that the spinal balance could be irreversibly affected and the scoliosis would progress rather than resolve. The prognostic rib-vertebral angle difference at the apex of the curve, as described by Mehta (1972), is produced by a combination of vertebral rotation and the action of gravity causing the ribs on the convexity to droop. The greater the tendency for the upper hemithorax to grow backwards, the greater the degree of vertebral rotation (and the rib-vertebral angle); the greater the rotation the more likely is the scoliosis to progress.

Why there should be such a marked decline in the number of children presenting in Edinburgh with infantile idiopathic scoliosis is not fully understood. In North America newborn children have always been laid prone and it has been suggested that this position could account for the much lower incidence of infantile

babies in restrictive blankets. However, the majority of babies are still not laid prone: only 25 per cent of 568 normal infants examined in their cots in Edinburgh had been laid prone and 75 per cent were supine (Brown *et al.* 1982). Seventy per cent of the babies who were laid supine showed a strong preference to turn towards the right. In my series, none of the infants who developed idiopathic scoliosis in the last six years had been laid prone. Wynne-Davies, in her series, found that before 1972 only three out of the 97 Edinburgh infants with scoliosis had been laid prone and all three had resolved spontaneously.

This study has shown a marked decline in the incidence of infantile idiopathic scoliosis in the community and it is therefore concluded that this is a preventable deformity. Evidence suggests that the onset of scoliosis may be associated with immobility in the supine or lateral position and it is possible that by laying the newborn infant in the prone position and encouraging much freer movement the incidence of this potentially severely crippling disorder may be further decreased.

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THE MANAGEMENT OF PROGRESSIVE INFANTILE IDIOPATHIC SCOLIOSIS

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Twenty-seven children with progressive infantile idiopathic scoliosis have been reviewed after long-term management. Twenty-two children had single thoracic curves which were diagnosed at an early age and treated in a modified Milwaukee brace until the age of ten years when the spine was corrected and fused. The mean correction after operation was 40 per cent of the initial degree of curvature seen in early childhood before treatment. Solid spinal fusion led to a further moderate loss in correction due to bending of the fusion mass before the spine became stable several years later. Five children had double structural curves and were treated only in a brace. This provided less satisfactory control of these curves but because of the minimal cosmetic deformity, extensive spinal fusion was avoided.

Infantile idiopathic scoliosis is a structural curve appearing without evident cause in a child before the age of three years (James 1951). Although rare in North America it is not uncommon in Great Britain; in a series of 200 consecutive patients presenting with idiopathic scoliosis at the Edinburgh Scoliosis Clinic between 1968 and 1972, eighty-two were of the infantile variety. Lloyd-Roberts and Pilcher (1965), in a study of 100 infants with infantile idiopathic scoliosis presenting in the first year of life, found that 90 per cent resolved spontaneously within two years; but in an earlier series of 212 children diagnosed before the age of three years and referred to a scoliosis clinic, only 77 per cent resolved (James, Lloyd-Roberts and Pilcher 1959). In the early stage both resolving and progressive types of infantile idiopathic scoliosis appear as virtually identical mild structural thoracic curves. However, with growth and if left untreated, progressive curves deteriorate to become one of the most severe forms of scoliosis (Figs. 1, 2 and 3). At skeletal maturity the majority of thoracic curves will measure over 100 degrees and the associated thoracic cage deformity results in cor pulmonale and probably in death in early adult life (Scott and Morgan 1955). A small minority of progressive thoracic curves that do not deteriorate so rapidly may later develop a second structural curve to the opposite side in the lumbar region; a very few patients may have both these curves present from the onset. The prognosis for these double structural curves is considerably better than that for a single thoracic curve.

The Edinburgh policy in the management of progressive infantile idiopathic scoliosis is to recognise the progressive curve early and start immediate conservative treatment. This management continues, if

possible, until the child is ten years old, when the spine is corrected and fused posteriorly. This policy applies to all patients with single thoracic curves but not necessarily to double structural curves in which the thoracic and lumbar components are usually well balanced and the cosmetic deformity is not great. Correction and fusion of double curves produces little cosmetic improvement and the resulting extensive spinal rigidity can be disabling. These curves are therefore managed conservatively until skeletal maturity unless the thoracic component deteriorates to the stage where future cardiothoracic complications become likely.

The purpose of this paper is to report the Edinburgh experience in the long-term management of progressive infantile idiopathic scoliosis diagnosed at an early age and treated by a combination of brace therapy and spinal fusion.

MATERIALS AND METHODS

A retrospective study has been made of twenty-seven children with progressive infantile idiopathic scoliosis who were first diagnosed at an early age at the Edinburgh Scoliosis Clinic and have undergone long-term treatment between 1964 and 1977. The many children who were treated elsewhere before being referred to Edinburgh at a later stage for further treatment have been excluded. In the earlier patients the diagnosis of a progressive as distinct from a resolving scoliosis was made by noting a radiological progression of the curve. Since 1968 a much earlier diagnosis has been made possible by estimating the rib-vertebra angle difference at the apex of the curve on the first spinal radiograph (Mehta 1972). Once the diagnosis of a progressive curve has been made, the likely prognosis and plan of treatment is discussed with the parents. The need for measures to prevent deterioration is emphasised and the child is fitted with a brace. The parents are informed that single thoracic curves will require correction and fusion at the age of ten years as part of the planned course of treatment and not because of the failure of the brace to control the deformity.

The brace which has been used in Edinburgh is a modification of

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Fig. 1

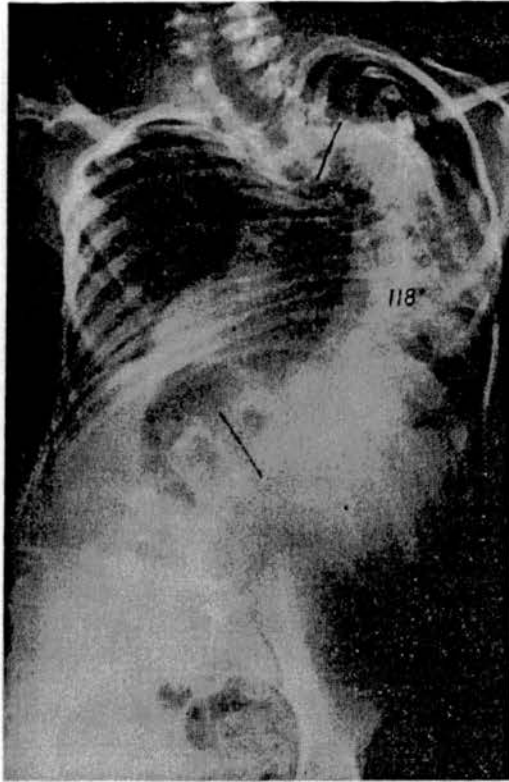


Fig. 2

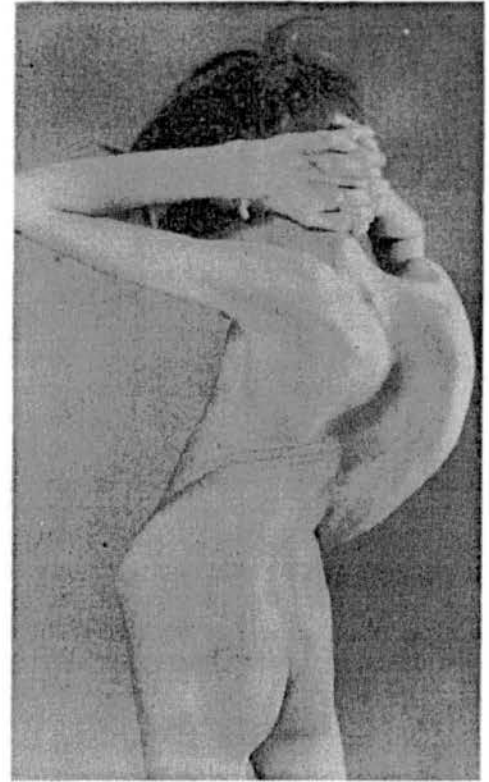


Fig. 3

Figure 1—The spinal radiograph of a one-year-old boy with a mild thoracic scoliosis. Figures 2 and 3—No treatment has been given and now at nine years old there is a severe thoracic scoliosis producing an extreme cosmetic deformity.

the Milwaukee brace (Blount, Schmidt and Bidwell 1958) and can be fitted as early as eighteen months of age once the child has developed a waist. If the curve is already greater than 40 degrees, preliminary correction in a Risser turnbuckle jacket is often carried out. The brace used in Edinburgh does not make use of the pads which have been advised by Blount and his associates as a means of applying a lateral corrective force to the site of greatest curvature. Instead, the Edinburgh brace relies only upon controlled distraction between the pelvic girdle and the mandible and occiput. The brace is worn day and night and is removed only for washing and bathing. Regular attendances are made at the Scoliosis Clinic where the brace is distracted and the progress of the curve is assessed radiographically.

Twenty-two children with single thoracic curves were reviewed; all had completed their treatment in a brace and had, at the age of ten years, undergone further correction in either a Risser localiser jacket or by Harrington instrumentation and posterior spinal fusion. Of the remaining five children with double structural curves, two were still being managed in a brace at the time of review, while the other three were skeletally mature and bracing had been discontinued. Erect spinal radiographs of each patient were measured using Cobb's method (Cobb 1960) and the rib-vertebra angle difference calculated from the earliest radiograph (Mehta 1972). Radiographs taken elsewhere before referral were often available and gave an indication of the rate of progression of the untreated curve.

RESULTS

Single thoracic curves

There were twelve boys and ten girls in this group with a mean rib-vertebra angle difference of 43 degrees (range 22 to 100 degrees) on the first spinal radiograph taken

before the age of three years. Mehta (1972) has suggested that rib-vertebra angle difference greater than 20 degrees is indicative of a progressive curve and all of the thoracic curves in this group were seen to progress. The curves extended from an upper limit of the fourth thoracic vertebra to a lower limit of the third lumbar vertebra and varied from seven to eleven vertebrae in length. Left-sided curves were twice as common as right-sided curves.

Single thoracic curves can be managed in three stages but the first stage was not carried out in thirteen patients.

Risser turnbuckle jacket. Nine patients, four boys and five girls, whose scoliosis when first seen was greater than 40 degrees, were corrected initially in a Risser turnbuckle jacket. The age at which the jacket was applied ranged from eighteen months to thirty months, and the mean initial curve of 62 degrees (range 40 to 90 degrees) was decreased by an average of 52 per cent to a mean curve of 29 degrees (range 15 to 40 degrees). After correction those infants who were too young to be fitted with an Edinburgh brace were managed temporarily in a plaster jacket.

Edinburgh brace. All twenty-two children were managed in the Edinburgh modification of the Milwaukee brace which was applied either initially or after preliminary correction in a Risser turnbuckle jacket.

After removal of the Risser jacket there was often some deterioration of the curve before the brace was applied. The mean age at bracing was four years three months (range one year nine months to eight years six months) and twelve out of the twenty-two children (55 per cent) were braced by their fourth birthday. The mean period in the brace was six years five months and 70 per cent of children wore the brace for more than five years.

Table I. Edinburgh brace treatment of infantile idiopathic progressive thoracic scoliosis (twenty-two patients)

Initial curve (degrees)	Number of patients	Brace correction	
		Best (per cent)	Final (per cent)
0-45	6	41	9
46-75	9	45	11
76-105	7	39	19

The mean initial curve just before bracing of all children with a single thoracic scoliosis was 63 degrees (range 29 to 105 degrees) and the mean percentage correction after bracing for one year was 38 per cent (Fig. 4). The mean best correction was 43 per cent and this was achieved by one year in eleven patients, by three years in eighteen patients and in all patients by five years. The greatest correction was obtained in those patients whose initial curves ranged from 46 to 75 degrees and the least correction in curves over 76 degrees (Table I). There was no significant correlation between the correction achieved and the length of the curve or the rib-vertebra angle difference.

After the initial best correction there was then a gradual deterioration of approximately one degree per year. The average progression of untreated single thoracic curves before referral to the Edinburgh Scoliosis Clinic was 12 degrees annually, although this figure varied from 6 to 20 degrees. After wearing the brace for five years the rate of deterioration became greater and the final mean correction in the brace at the age of ten years just before spinal fusion was 11 per cent (Fig. 4); this represents a mean angle of curvature at the end of bracing of 55 degrees (range 20 to 78 degrees). The larger initial curves, however, maintained a better percentage correction than lesser curves (Table I).

Spinal fusion. All single thoracic curves were routinely fused at the age of ten years. In the first fifteen patients correction in a Risser-type of localiser cast and fusion resulted in a mean correction of 40 per cent; this represented an improvement from a mean angle of 71 degrees out of the brace before operation to 42 degrees afterwards. A greater mean correction of 53 per cent was achieved in the last seven patients by using Harrington instrumentation: a mean improvement in the curve from 62 degrees out of the brace before operation to 30 degrees afterwards.

After operation the degree of curvature in twenty out of twenty-two patients was less than that measured on the radiograph taken in infancy and before the application of the Risser turnbuckle jacket or Edinburgh brace. The two patients whose curves were still slightly greater after operation had both been corrected in a Risser localiser jacket. The mean correction after operation for the twenty-two patients was 40 per cent of the degree of curvature when first seen in early childhood before treatment (Fig. 4).

Of the fifteen patients whose curves were treated in a localiser cast three were later found to have pseudarthroses which were repaired at periods varying from nine to thirty-three months after the attempted fusion. All patients have now been followed for an average of seven years (range three to ten years) and in those patients with a solid fusion the mean loss in correction after one year was 6 degrees and at final follow-up was 17 degrees. In the three patients with pseudarthroses the mean loss in correction after one year was 3 degrees and at final follow-up was 27 degrees.

The seven patients whose curves have been corrected by means of Harrington instrumentation have

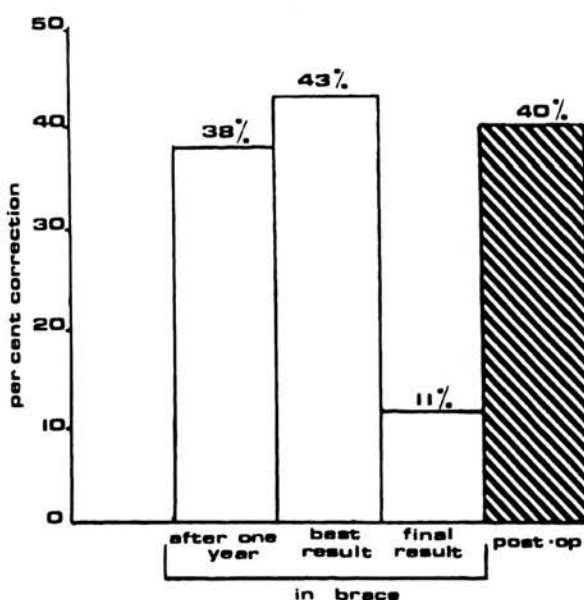


Fig. 4

A histogram showing the mean degree of correction obtained during brace therapy and after spinal fusion of twenty-two single thoracic curves as compared with the degree of curvature when first seen in early childhood and before treatment.

been followed for an average of twenty-one months (range fifteen to forty-two months). All have been surgically re-explored at six months and no pseudarthroses found. The mean loss in correction at one year was 5 degrees and at final follow-up there had been no further loss.



Fig. 5



Fig. 10

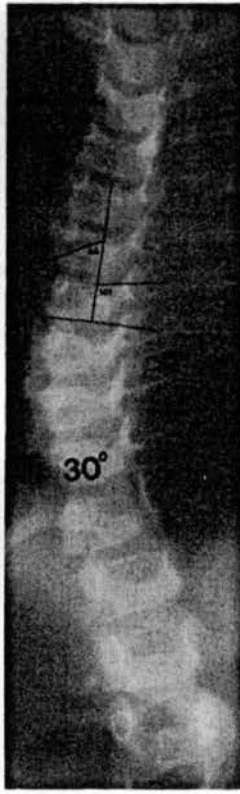


Fig. 6

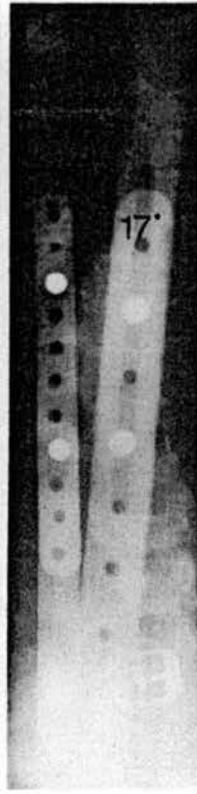


Fig. 7

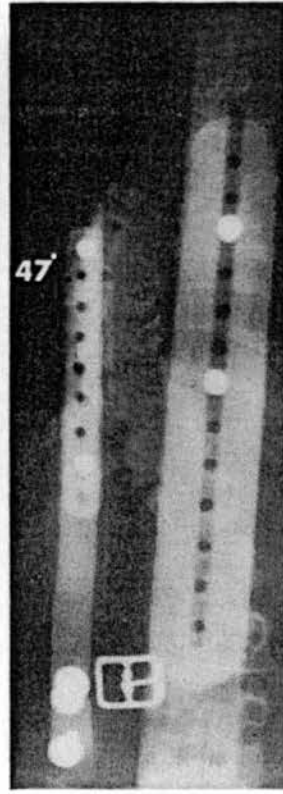


Fig. 8

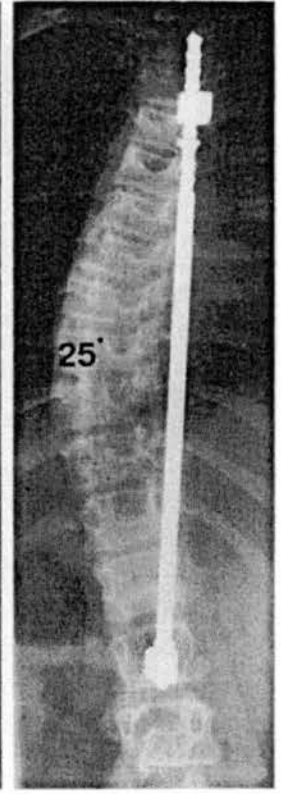


Fig. 9

Case 1. Figure 5—A boy aged two years and ten months with a moderate left thoracic scoliosis when first seen. Figure 6—The spinal radiograph shows a positive rib-vertebra angle difference of 37 degrees indicating a probable progressive curve. Figure 7—The best correction in the brace was achieved after three years. Figure 8—After seven years of brace therapy and just before operation. Figure 9—One year after Harrington rod correction and spinal fusion the degree of spinal curvature is better than when first seen. Figure 10—There has been a good cosmetic result.

The following two case reports illustrate the treatment and results obtained in two patients, one with a mild and the other with a severe thoracic scoliosis.

Case 1. This boy's mother first noticed that his spine was not straight when he was a few months old. She was reassured by her local doctor but became dissatisfied when at the age of two years and ten months she noted that the deformity had increased. The child was then seen at the Edinburgh Scoliosis Clinic where he was found to be normal except for a mild left thoracic scoliosis with an associated rib hump (Fig. 5) and left plagiocephaly. Radiographs revealed a left thoracic scoliosis extending from the fourth thoracic vertebra to the first lumbar vertebra and measuring 30 degrees with a rib-vertebra angle difference of 37 degrees (Fig. 6).

The child was immediately fitted with an Edinburgh brace which he wore until he was nearly ten years old. The best correction in the brace was achieved at the age of five years and nine months when the curve measured 17 degrees (Fig. 7). Good correction was maintained until the age of eight years and five months when the curve began slowly to deteriorate. At the age of nine years and seven months, when the curve measured 47 degrees in the brace (Fig. 8), a posterior spinal fusion with Harrington instrumentation was performed. The curve measured 51 degrees out of the brace before operation and was corrected to 20 degrees without complication. The child has now been followed for over one year with only a 5 degree loss in correction (Figs 9 and 10).

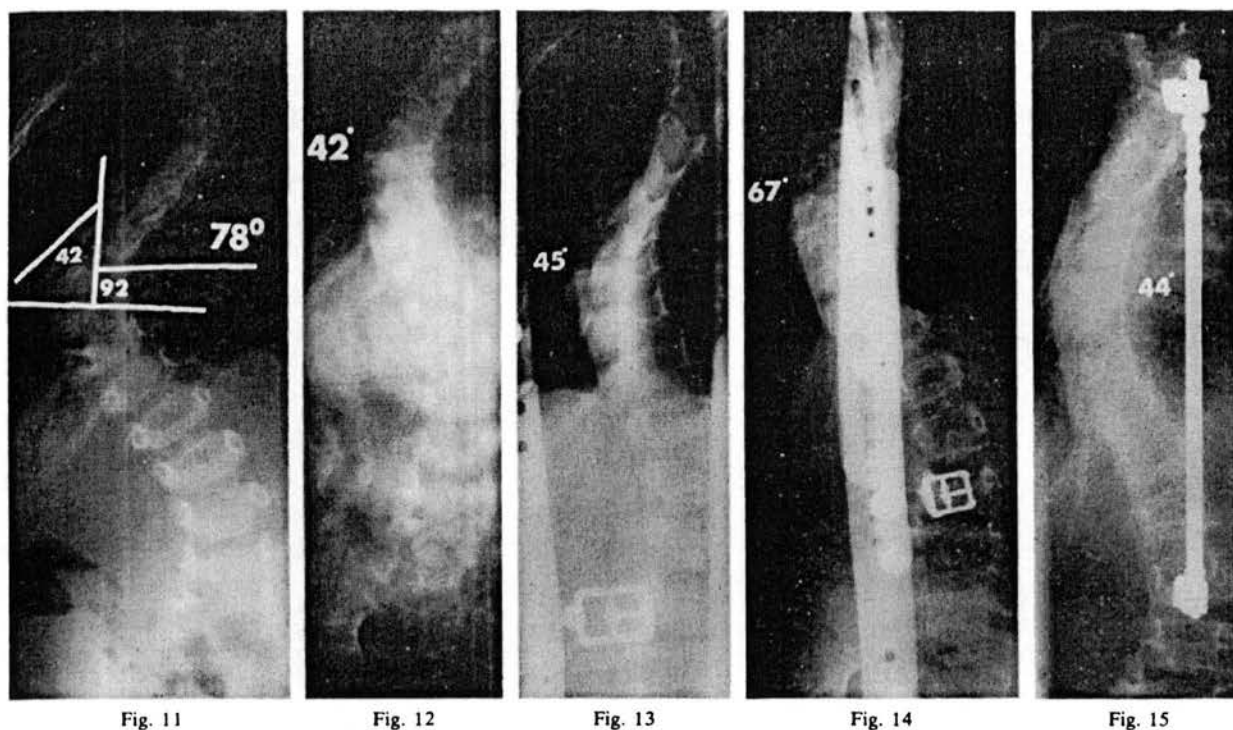
Case 2. This girl was first seen at the Edinburgh Scoliosis Clinic at the age of two years. She had been delivered by Caesarean section and had developed normally until the age of eighteen months when a rib hump

was first noted by her mother. On examination the infant was found to be normal except for a severe left thoracic scoliosis with an associated prominent left rib hump and a left-sided plagiocephaly. Radiographs revealed a left thoracic scoliosis which extended from the fifth thoracic vertebra to the first lumbar vertebra and measured 78 degrees. The rib-vertebra angle difference was 50 degrees (Fig. 11).

She was admitted to hospital where a Risser turnbuckle jacket was applied and the deformity corrected to 35 degrees. Two months later a moulded plaster jacket was applied (Fig. 12) and maintained until the age of two years and eight months when she was fitted with an Edinburgh brace. The best correction in the brace was achieved at the age of four years and eleven months when the curve measured 45 degrees (Fig. 13). Good correction was maintained until she was aged eight years six months when the curve slowly began to deteriorate. At the age of ten years when the curve measured 67 degrees in the brace (Fig. 14) she underwent a posterior spinal fusion with Harrington instrumentation. Before operation her curve measured 84 degrees out of the brace and this was corrected to 41 degrees without complication. The patient has now been followed for over one year with only a 3 degree loss in correction (Fig. 15).

Double curves

There were three boys and two girls in this group. In all cases the rib-vertebra angle at the apex of the convexity of the thoracic curve was greater than that on the concavity so producing a negative rib-vertebra angle difference; this ranged from -9 to -50 degrees (mean



Case 2. Figure 11—The first spinal radiograph at the age of two years shows a severe left thoracic scoliosis with a positive rib-vertebra angle difference of 50 degrees indicating a probable progressive curve. Figure 12—After correction in a Risser turnbuckle jacket and application of a holding plaster jacket. Figure 13—The best correction in the brace was achieved after two years. Figure 14—After seven years and eight months of brace therapy and just before operation. Figure 15—One year after Harrington rod correction and spinal fusion the degree of spinal curvature is better than when first seen.

–22 degrees). Mehta (1972) found that a negative rib-vertebra angle difference at the apex of the thoracic curve was often indicative of a second structural curve in the lumbar region and this proved so in all these patients. The upper curves extended from the sixth to the twelfth thoracic vertebra and the lower curves from the tenth thoracic to the fifth lumbar vertebra. The mean number of vertebrae in each curve was six. There were three patients with left thoracic and right lumbar curves and two with the reverse pattern. The mean initial thoracic curve measured 45 degrees (range 34 to 55 degrees) and the mean initial lumbar curve measured 48 degrees (range 41 to 60 degrees).

The rate of deterioration of these double structural curves was much less than in the single thoracic curves with the result that bracing was not necessary until a later age. The brace was applied at a mean age of seven years and nine months (range four to thirteen years) and was worn for a mean period of three and a half years.

The mean best correction was achieved by two years in all patients: 14 per cent for thoracic curves and 23 per cent for lumbar curves. Three of these children have now reached skeletal maturity and are out of the brace. Two children are still wearing the brace after eighteen and forty-four months. The mean correction for the thoracic curves when last seen was –1 per cent (range –26 per cent to +17 per cent) and for the lumbar curves 17 per cent (range 2 to 39 per cent).

Case 3. This boy was first seen at the age of four years and seven months. A few months after birth, his mother noticed a slight prominence of one side of his back posteriorly and also an asymmetry of his face but the spinal deformity did not appear to deteriorate until just before his referral to the Scoliosis Clinic. On examination he was found to be normal except for a mild left thoracic scoliosis and mild right lumbar scoliosis both of which showed clinical rotation. Radiographs confirmed a double structural scoliosis with a left thoracic curve extending from the seventh to the twelfth thoracic vertebra measuring 28 degrees and a lumbar curve from the twelfth thoracic to the fifth lumbar vertebra measuring 22 degrees. The rib-vertebra angle difference was –40 degrees (Fig. 16).

No specific treatment was prescribed and the child was seen regularly at the Scoliosis Clinic where the progress of the curve was followed radiographically. There was little deterioration until the age of eight years and eight months when the thoracic curve was found to be 45 degrees and the lumbar curve 41 degrees (Fig. 17). Although the cosmetic deformity was small it was decided to treat the curves in an Edinburgh brace to prevent further deterioration. The best correction in the brace was achieved at the age of ten years and six months when the thoracic curve measured 30 degrees and the lumbar curve 24 degrees (Fig. 18). At present he is still wearing his brace at the age of fourteen years and there has been a slight deterioration of the thoracic curve to 39 degrees and the lumbar curve to 36 degrees (Fig. 19). The cosmetic deformity remains small and it is proposed to continue to use the brace until skeletal maturity if possible.

Complications

Clinically significant orthodontic moulding occurred in eight of the twenty-two patients with single thoracic curves (36 per cent) and in one of the five patients with double structural curves but after final removal of the

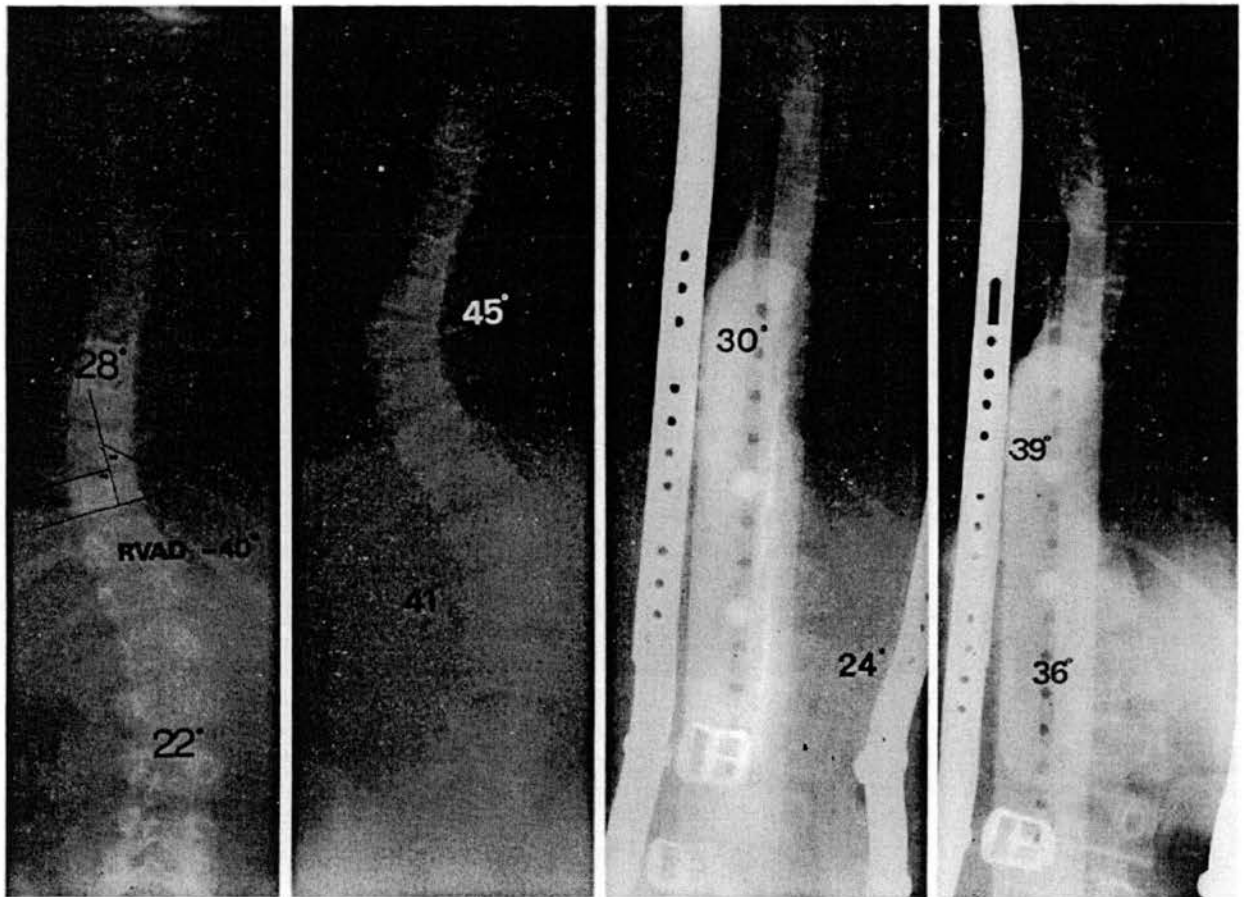


Fig. 16

Fig. 17

Fig. 18

Fig. 19

Case 3. Figure 16—A negative rib-vertebra angle difference of 40 degrees at four years seven months and the obliquity of the twelfth rib on the concavity of the thoracic curve indicate a probable double structural curve. Figure 17—By eight years and eight months the double structural curves had become more apparent and a brace was applied. Figure 18—The best correction in the brace was achieved after two years. Figure 19—After five years of brace therapy both curves have deteriorated slightly but are still better than before the brace was applied.

brace and with further growth the deformity usually resolved. Pressure sores developed in four patients but responded to removal of the brace for a few days and adjustment of the pelvic girdle.

DISCUSSION

Posterior fusion is a well-established method of preventing an increase in spinal deformity but unfortunately cannot be applied to very young children. The fused area will not grow in length and will therefore cause a relative shortening of the spine as the child grows and also a possible increasing lordosis due to continuing growth of the unfused vertebral bodies anteriorly (Ponseti and Friedman 1950). In order to minimise these difficulties James (1967) recommended that posterior spinal fusion be postponed until the age of ten years unless progression of the curve could not be controlled by other means. A long period of conservative treatment is therefore necessary to prevent deterioration of a curve which develops before the age of three years and can only be safely fused after the age of ten

years. The most effective means of preventing deterioration is the Milwaukee brace, although if the curve is already severe correction in a Risser turnbuckle jacket may be necessary before applying the brace. The Edinburgh brace is a modification of the Milwaukee brace and depends only on distraction. The modern Milwaukee brace relies less on distraction and more on an axially directed force applied through the ribs to the site of maximum spinal curvature. This is a dynamic force produced by the patient who performs specific exercises against a fixed pad applied over the rib hump. In curves over 40 degrees the axially directed force becomes progressively less effective and distraction becomes more important as the size of the curve increases (Blount and Moe 1973).

In our study many patients were referred late for treatment and the mean degree of curvature for a single thoracic scoliosis immediately before bracing was 63 degrees. The Edinburgh brace produced a mean best correction of 43 per cent in the twenty-two patients with single thoracic curves (Fig. 4) and the greatest percentage correction was obtained in curves measuring

46 to 75 degrees (Table I). The brace, however, was much less effective in gaining correction in the five patients with double structural curves: a mean best correction of 14 per cent for thoracic curves and 23 per cent for lumbar curves.

The best correction of single thoracic curves was obtained in 59 per cent of our patients in two years, but some required a five-year period of bracing. After the initial best correction there was then a slow deterioration of approximately one degree per year, but after five years in the brace the rate of deterioration usually increased and by the age of ten years the overall mean curve correction was 11 per cent. However, larger thoracic curves maintained better correction than lesser curves as might be expected in a pure distraction system. The final degree of correction of single thoracic curves in our patients was less than that achieved at skeletal maturity in juvenile and adolescent idiopathic scoliosis by Moe and Kettleson (24 per cent) and by Keiser and Shufflebarger (26 per cent) but our infantile patients wore their brace on average for twice the period reported in these series.

In Edinburgh it has been the policy to carry out a posterior spinal fusion of all progressive single thoracic curves as a routine procedure at the age of ten years. If spinal fusion were not performed a brace would have to be worn until skeletal maturity which in boys can be as late as eighteen years. By ten years of age 70 per cent of our patients had already worn their brace for over five years and we consider it unfair to subject these children to an even longer period in the brace unless it is absolutely necessary. Immediately after spinal fusion the degree of curvature in all but two patients was better than that before the onset of conservative treatment in early childhood, the overall degree of correction being 40 per cent (Fig. 4). Harrington instrumentation is now

routinely used in nearly all curves and had it been applied to these two patients the degree of correction might have been greater. Unfortunately in these young children the development of solid spinal fusion did not necessarily prevent further moderate loss in correction. Those patients with a solid spinal fusion whose scoliosis had been corrected in a localiser cast and who have been followed for an average of seven years after operation have lost an average of 17 degrees of correction. Nearly all of this loss occurred in the first two to three years and is thought to be due to bending of the solid but immature fusion mass before it eventually became stable. Those patients whose spines were corrected by means of Harrington instrumentation all developed a solid spinal fusion, and in an effort to produce a stronger fusion mass all received a massive onlay of autogenous cancellous bone strips after deep decortication at the time of the routine interfacetal fusion. These patients have now been followed for a maximum of three and a half years and there has so far been no loss in the mean correction after the first year. A much longer follow-up is necessary before any definite conclusion can be drawn.

In the much smaller number of children with double structural curves the possible disability resulting from a nearly total spinal fusion at a relatively early age outweighs the advantages, and brace therapy is therefore continued until skeletal maturity. Fortunately even quite large double structural curves are cosmetically acceptable, but should the thoracic curve continue to progress to the stage where cardiorespiratory complications seem likely, Harrington instrumentation and posterior spinal fusion are performed immediately.

We therefore suggest that a planned course of treatment incorporating both brace therapy and spinal fusion is an effective means of managing progressive infantile idiopathic scoliosis.

We would like to thank Professor J. I. P. James for allowing us to review his patients and also Mr M. Devlin for his photographic assistance.

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THE PEDIATRIC SPINE

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16 INFANTILE IDIOPATHIC SCOLIOSIS

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Infantile idiopathic scoliosis is a structural curve that appears without evident clinical or radiologic cause in the first 3 years of life. In these children there is no evidence of an underlying neurologic or muscular disorder. The spinal radiograph shows a curvature, but apart from this the vertebrae are normal and there are no developmental abnormalities, such as are found in congenital scoliosis.

The first description of what we now recognize to be infantile idiopathic scoliosis was by Harrenstein in 1936.⁵ It was not, however, until 1951 that James⁹ provided a much more full description of this type of scoliosis and classified it as being "infantile" because it occurred at an early age and "idiopathic" because there was no obvious cause. He also noted that infantile idiopathic scoliosis had a different pattern and prognosis from idiopathic scoliosis occurring in older children.¹⁰ Infantile curves were more common in boys and the majority had a left thoracic scoliosis. After the age of 3 years, there was a gradual change in the clinical pattern until by 10 years the majority of patients developing adolescent idiopathic scoliosis were girls and a high proportion had a right thoracic curve. The most striking difference, however, was that the majority of infantile curves disappeared spontaneously, whereas adolescent and juvenile curves did not disappear and nearly all progressed.^{10,22} In 1959, James and associates¹¹ used the terms "resolving" and "progressive" to differentiate between these two types of infantile idiopathic scoliosis. They noted that progressive curves developed rapidly and relentlessly, becoming one of the most severe forms of scoliosis, whereas resolving curves disappeared completely within a few years and did not return.

INCIDENCE

Infantile idiopathic scoliosis has been relatively common in Great Britain, but rare in North America. In a study of 157 consecutive

patients with idiopathic scoliosis seen in Edinburgh between 1968 and 1971, 41% had infantile curves, 7% had juvenile curves, and 52% had adolescent curves. However, since that time, there has been a gradual change in the relative frequencies of the different types of idiopathic scoliosis.¹⁷ Of 153 patients with idiopathic scoliosis seen at the same clinic in the years 1980 to 1983, only 4% had infantile curves, 7% had juvenile curves, and 89% had adolescent curves. The relative frequencies of the different types of idiopathic scoliosis seen in Edinburgh now approaches that found in North America. A survey in Boston of 208 patients found that 0.5% had infantile curves, 10.5% had juvenile curves, and 89% had adolescent curves²⁴ (Fig. 16-1). A similar decline in the previously high incidence of infantile idiopathic scoliosis has been observed by Mau in Tübingen, West Germany.^{13,14}

ETIOLOGY

The exact cause of infantile idiopathic scoliosis is unknown, but it is thought to be multifactorial in origin. Wynne-Davies^{30,31} studied the familial aspect of the condition in Edinburgh and found that there is a genetic tendency to develop the deformity, but this by itself is insufficient and requires to be triggered by other adverse factors that may be medical or environmental. These adverse factors must, however, act mainly after birth because the scoliosis is rarely, if ever, present at birth. The balance in each infant is likely to be different and an infant with a strong genetic tendency requires little triggering action, and vice versa.

The mysterious fact that the condition is rare in North America and relatively common in Great Britain cannot be due to any genetic difference in the two white populations. There may be some beneficial aspect of the environment of North America that is actively preventing the tendency to develop the deformity, or

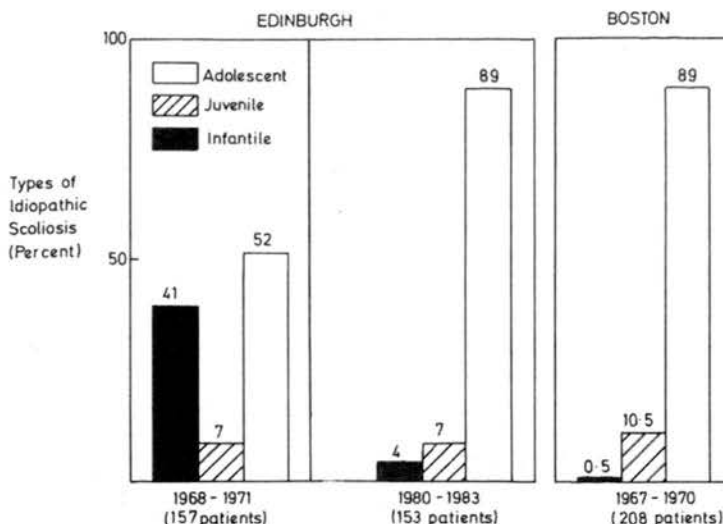


Figure 16-1. The relative frequencies of infantile, juvenile and adolescent idiopathic scoliosis in Edinburgh and Boston. (Reproduced with permission from McMaster.¹⁷)

there may be some adverse triggering factor present in Great Britain.

A clue to this factor is found in the very close association between infantile idiopathic scoliosis and plagiocephaly. Plagiocephaly is a plastic deformation of the skull that occurs when an immobile infant habitually lies toward one side and the action of gravity deforms the immature plastic skull. The uppermost side of the face and head flows backward, becoming recessed, while the lower ear, which is in contact with the mattress, is pushed forward, producing the commonly associated bat ear (Figs. 16-2 and 16-3). This deformity, which is not usually present at birth, was found in 28% of 223 normal Edinburgh infants examined between the ages of 6 weeks and 6 months and the recessed side of the head was on the left in 83%,^{6,31} left-sided plagiocephaly (Fig. 16-4A and B). Premature babies have an even higher incidence of plagiocephaly (63%) and this is thought to be due to their greater immobility.¹ The plagiocephaly, however, rarely persists and once the child becomes mobile, it usually disappears by the age of 6 years.

In a study of 144 children with infantile

idiopathic scoliosis seen in Edinburgh between 1968 and 1983, plagiocephaly was present in 96% of those examined in the first 6 months of life.¹⁷ The plagiocephaly was equally prevalent in both resolving and progressive curves, and the recessed side of the head was on the left in 81%. The scoliosis in these infants was rarely noted at birth, but, like the plagiocephaly, developed within the first 6 months of life in 70%. The convex side of the curvature corresponded with the side of the plagiocephaly in all except four infants with resolving curves. This close association between the presence, time of onset, and side of the deformity of both plagiocephaly and infantile idiopathic scoliosis suggests a common pathogenesis, with posture being a factor. Wynne-Davies³¹ also found that in infants with idiopathic scoliosis there was a greater than normal incidence of prematurity, low birth

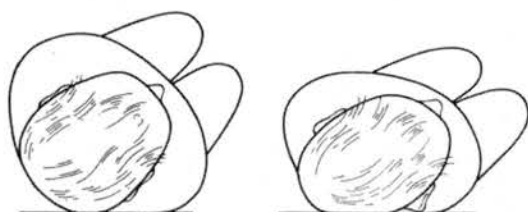


Figure 16-2. The mechanism of postural moulding of the head and thorax. (Reproduced with permission from McMaster.¹⁷)

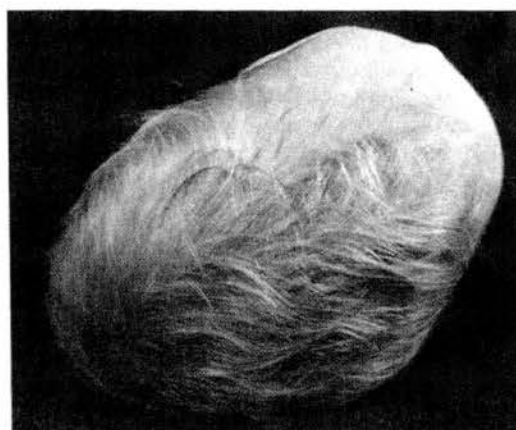


Figure 16-3. Postural moulding of the head, or plagiocephaly. (Reproduced with permission from McMaster.¹⁷)

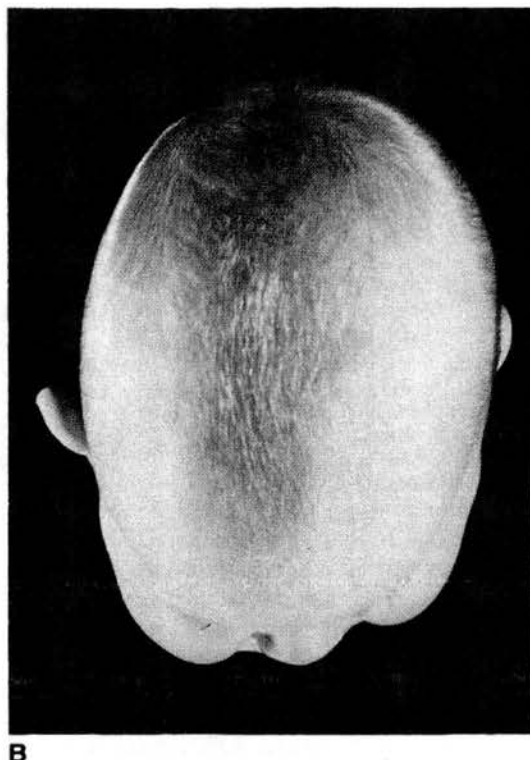


Figure 16-4. A and B. Left-sided plagiocephaly with contralateral bat ear. (Reproduced with permission from McMaster.¹⁷)

weight, mental retardation in males, older mothers, poorer homes, congenital heart disease, and delayed maturation of muscles and ligaments, the latter being evident from hernias and congenital dislocation of the hip. All of these factors contribute to immobility of the newborn and, when combined with posture, could trigger the development of a scoliosis in a genetically predisposed infant.

Observations in Edinburgh¹ and elsewhere¹³ have shown that during the first few months of life, there is a natural tendency for all newborn infants when laid supine to turn partially toward their right side, and this would account for the predominance of left-sided plagiocephaly in both normal infants and those with scoliosis. In my opinion, when an immobile infant is laid supine and partly turns toward the right side, the immature thorax also undergoes plastic deformation due to the influence of gravity. The uppermost hemithorax, like the skull, tends to flow backward, causing the thoracic vertebrae to rotate backward on their long axis, (Figs. 16-2 and 16-5). There is a biomechanical coupling of axial rotation and lateral bending of the spine; axial rotation of the mid and lower thoracic vertebrae can on occasion induce a lateral curvature in which the anterior aspects of the verte-

bral bodies point toward the convexity of the curve.²⁸ Backward vertebral rotation in the infant could therefore induce an upward curvature of the spine and a left-sided thoracic scoliosis in infants lying toward their right side. This tendency to upward curvature is also increased by pressure from the mattress on the ribs of the lower hemithorax. Once the central nervous system and the vertebral musculature matures, the infant becomes much more mobile and the tendency to spinal curvature is usually overcome, accounting for the high incidence of resolving curves. If, however, there is marked vertebral rotation associated with the genetic, medical, and environmental factors discussed earlier, it is possible that the spinal balance could be irreversibly tipped and the scoliosis would progress rather than resolve.

The prone-lying position is theoretically advantageous because it prevents asymmetrical moulding of the thorax and promotes early maturation of the spinal extensor muscles. In North America, newborn infants have always been laid prone and allowed to move freely, and this could account for the much lower incidence of infantile idiopathic scoliosis. In the past, infants in Great Britain were usually laid supine or toward one side and wrapped in blankets. However, over

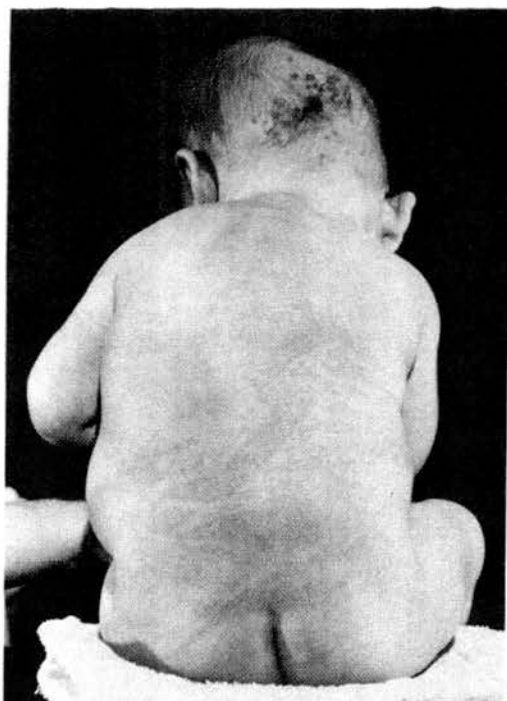


Figure 16-5. A 4-month old boy with a left thoracic infantile idiopathic scoliosis. He had habitually been laid supine and partially turned toward his right side, resulting in postural molding of his head and trunk. The creases on the right side of his trunk indicate his relative immobility. The head shows left-sided plagiocephaly, and there is a right bat ear that has been against the mattress. (Reproduced with permission from McMaster.¹⁷)

the last decade, there has been an increasing tendency toward the prone position and because of more frequent central heating the infants are less restricted by blankets. Could this account for the decreased incidence of the condition seen in Edinburgh?¹⁷

CLINICAL PRESENTATION

Boys are more commonly affected than girls, in a ratio of 3 to 2, and the deformity is usually first noticed by the mother when she lays the naked child prone across her knee. In this position the small rib hump on the convexity of the curve becomes more apparent, although the curve itself may not be obvious in a chubby infant (Fig. 16-5). When first seen at an early stage, progressive and resolving curves appear indistinguishable both clinically and radiologically. A single structural curve in the thoracic region is by far the most common and is nearly always convex to the left. Thoracolumbar curves are much less common and lumbar curves are

rare. Double structural curves also occur but are less frequent than single curves. In a study of 144 consecutive children with infantile idiopathic scoliosis seen in Edinburgh, 138 had single curves (96%) and only six had double curves (4%).¹⁷ Of the single curves, 130 were thoracic, seven were thoracolumbar, and there was only one lumbar curve. Eighty-two percent of the single curves were convex to the left. These findings are similar to those found in other series.

Congenital dislocation of the hip has also been found to occur more frequently in association with infantile idiopathic scoliosis. A dislocated or dislocatable hip was found in 6.4% of the Edinburgh infants and in unilateral hip disorders; the convexity of the thoracic scoliosis was on the same side as the dislocation.⁸ Ceballos and associates³ found a 24.8% incidence of hip dysplasia, but could not relate this to the side of the curvature.

NATURAL HISTORY

The reported incidence of infantile idiopathic curves that resolve has varied from 52 to 92%.^{3,4,12,27,31} In Edinburgh 74% of the infantile curves resolved and 26% progressed.¹⁷ However, the majority of these reports come from specialized centers for the treatment of scoliosis and may therefore reflect a bias toward progressive curves. The true incidence of resolving curves is probably nearer to 90%.

RESOLVING SCOLIOSIS

Resolving curves are nearly always in the thoracic region (95%) and much less frequently in the thoracolumbar region (5%). Rarely, if ever, are double structural curves seen to resolve. Resolving curves tend to be long and gentle, with a maximum angle of between 10° and 20° in slightly more than 50%. Very few resolving curves are greater than 30° and by far the largest curve reported to disappear was 46°.⁴ However, it should be remembered that although the majority of these curves are small, they are structural and do not fully correct on suspension.

The majority of resolving curves disappear completely by the age of 1 or 2 years, and no fully resolved curve has been known to reappear. However, a few curves resolve more slowly and do not fully correct until 7 or 8 years of age. In these patients the lateral curvature is often first to disappear, leaving vertebral rotation to resolve more slowly over a period of several years

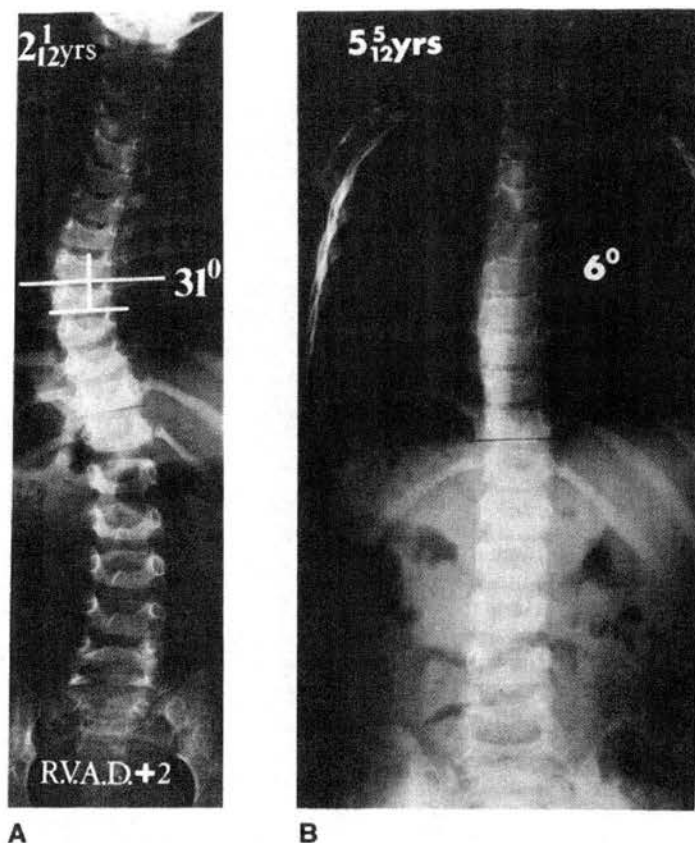


Figure 16-6. **A.** A boy, aged 2 years 1 month old, with a 31° left thoracic scoliosis. The rib-vertebral angle difference is plus 2° indicating that the scoliosis should resolve. **B.** No treatment was given and by the age of 5 years, 5 months, the scoliosis had virtually resolved, but the vertebral rotation remains and is the last to disappear.

(Fig. 16-6A and B). This pattern of resolving suggests that the vertebral rotation is the primary deformity.

PROGRESSIVE SCOLIOSIS

Thoracic Curve

The most common progressive infantile idiopathic curve occurs in the thoracic region (75 to 80%) and is usually more than 20° when first seen (Fig. 16-7A). At first, these are normal, healthy, chubby infants, but as the scoliosis rapidly increases, they soon cease to thrive. By the age of 6 years, the child has become thin and dwarfed, the face is often sharp and pinched, and there is a general lack of subcutaneous fat. The spinal curvature, which becomes very severe, usually extends from T5 to T12, and there is marked vertebral rotation with a large rib hump and distortion of the thoracic cage (Fig. 16-7B to F). The trunk is shortened and there is a characteristic humpedback appearance, which is due to a combination of vertebral rotation and a kyphosis in the upper thoracic region.¹¹ The presence of a kyphosis in infantile idiopathic sco-

liosis is very different from adolescent idiopathic scoliosis, which is more often associated with a lordosis.

Although the majority of progressive thoracic curves deteriorate rapidly, there are a few that progress more slowly. In contrast to the more rapidly progressive curves, these children maintain a relatively normal physical appearance for their age. Their curves are more flexible in the early stages and do not develop the severe vertebral rotation and apparent kyphosis seen in the rapidly progressive curves. Mehta¹⁹ has described these two types of progressive scoliosis as benign and malignant.

Double Structural Curves

Double structural curves are nearly always progressive and account for 10 to 15% of all progressive curves. They occur on opposite sides in the thoracic and lumbar regions and are equally distributed to either side of the spine. The thoracic curve usually extends from the T1 to T11 and the lumbar curve from the T11 to L4. This double pattern of curvature presents at a later age than single progressive thoracic curves. A common presentation of a double structural sco-

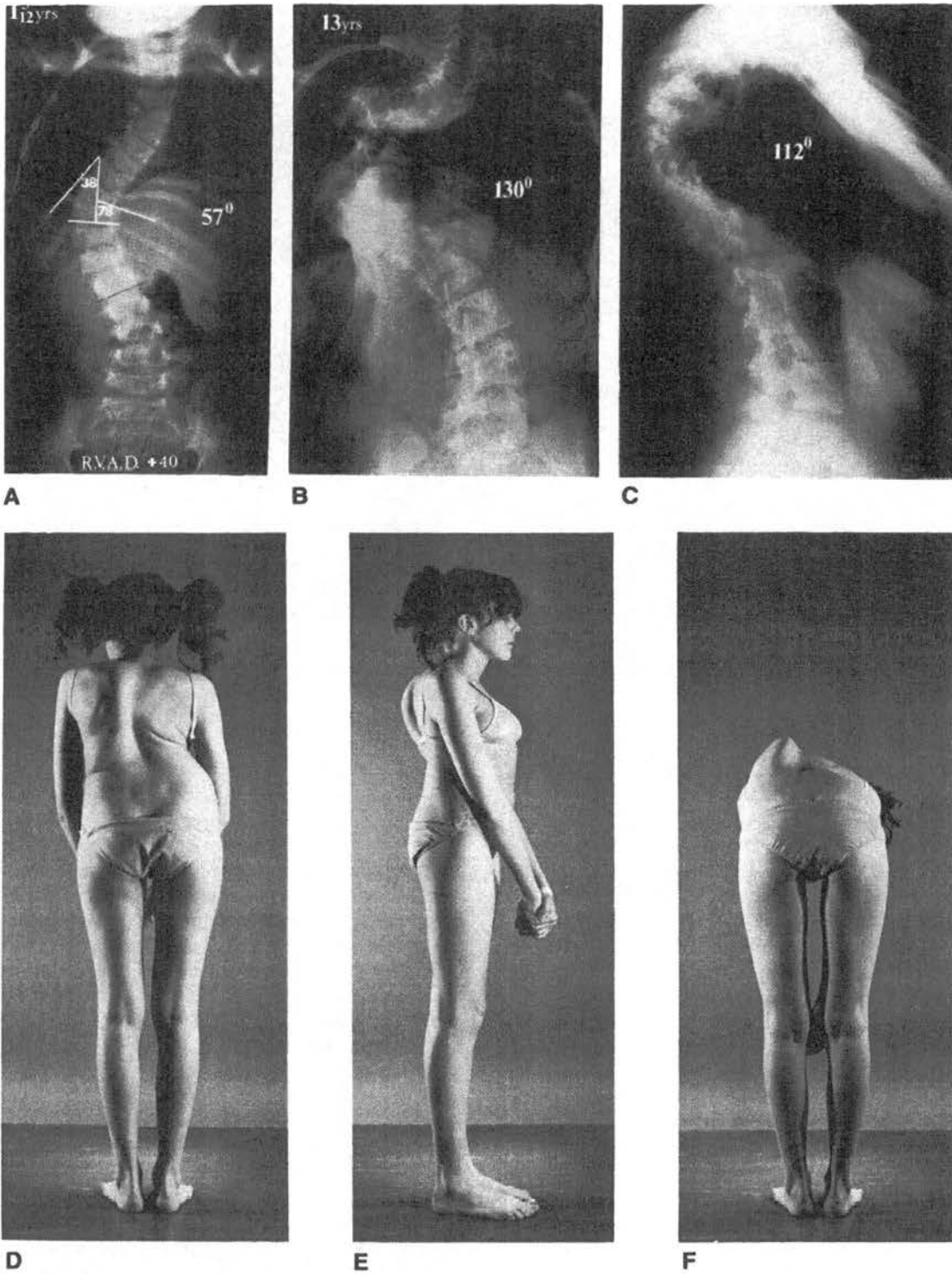


Figure 16-7. **A.** A girl, 1 year 5 months, with a 57° left thoracic scoliosis. The rib-vertebral angle difference is +40°, indicating that the scoliosis should progress. **B** and **C.** No treatment was given and by the age of 13 years the scoliosis progressed to 130° and there was an upper thoracic kyphosis of 112°. This is a malignant progressive curve. **D, E,** and **F.** Clinically there is a very severe deformity with marked distortion of the chest, a large rib hump, and an apparent kyphosis in upper thoracic region. The trunk is shortened and the child is dwarfed.

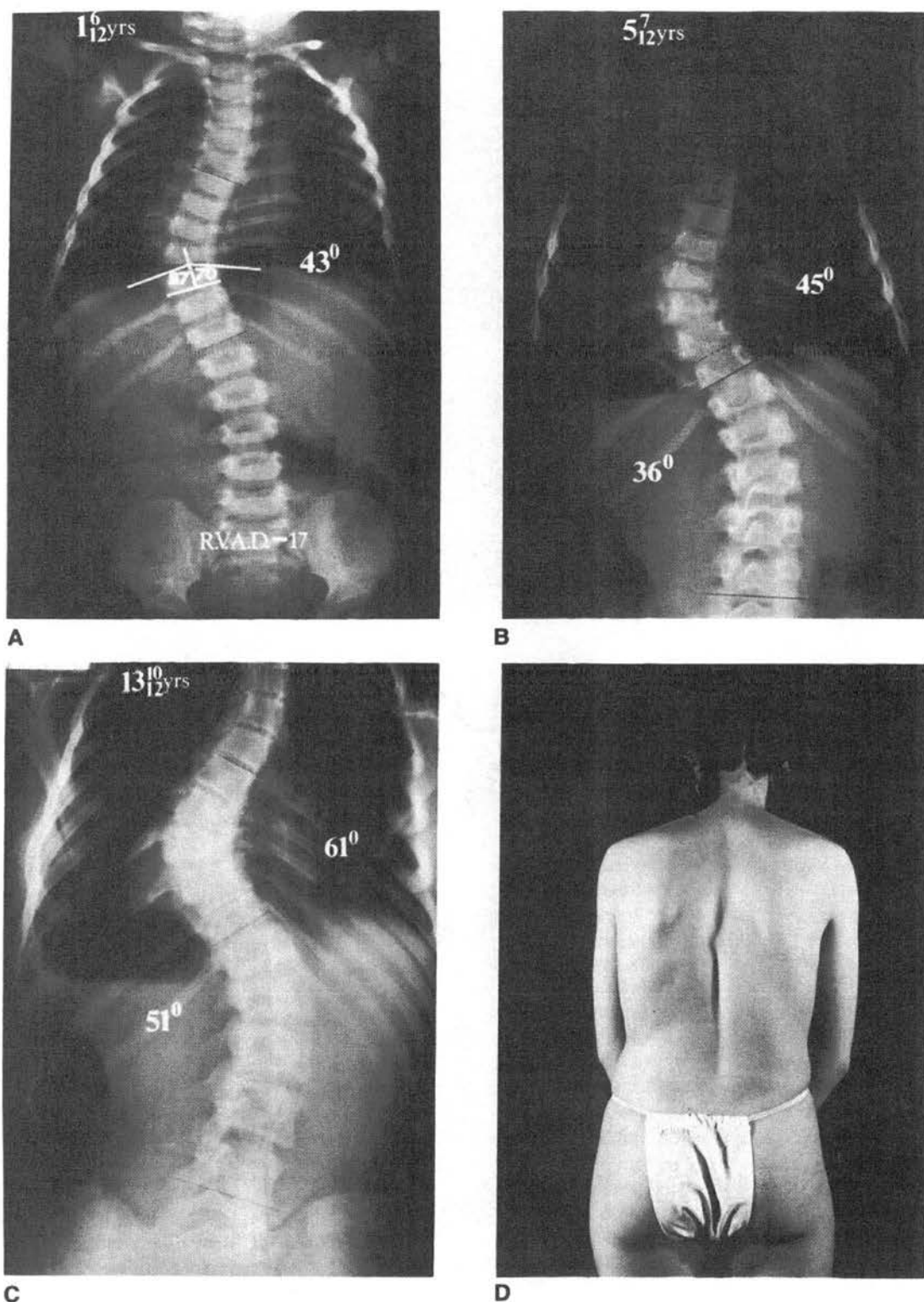


Figure 16-8. **A.** A girl, 1 year, 6 months with a 43° left thoracic scoliosis. The rib-vertebral angle difference is -17°, indicating that the thoracic curve could be one component of a double structural scoliosis and the lumbar curve is yet to appear. **B.** No treatment was given and by the age of 5 years, 7 months, the lumbar vertebrae rotated in the opposite direction to the thoracic vertebrae and a lumbar scoliosis is developing. **C.** At the age of 13 years 10 months, a double structural scoliosis is present, affecting the thoracic and lumbar regions. **D.** Clinically, the curves are well balanced and there is only a mild cosmetic deformity.

liosis is for the thoracic curve to appear first in the second or third year of life (Fig. 16-8A) and then to progress less rapidly than expected for a single thoracic curve. It is only in the next few years that a second structural curve becomes apparent on the opposite side in the lumbar region (Fig. 16-8B). Although these curves progress less rapidly than single thoracic curves, they still have a poor prognosis because of the long period of growth before skeletal maturity. The double curves, however, remain balanced and do not produce the same severity of cosmetic deformity that is seen with single thoracic curves (Fig. 16-8C and D).

Thoracolumbar and Lumbar Curves

These curves are much less frequent and do not progress as rapidly as the thoracic curves, but they have a worse prognosis than similar curves developing in adolescence.

Untreated Curves

Nowadays no progressive infantile idiopathic curve is left untreated and we do not see the natural history of the condition. However, James and associates¹¹ were able to study progressive curves in 107 children, who, because of World War II, did not receive treatment until a late stage.¹¹ They attempted to establish the natural history by noting the age and the greatest angle of curvature before treatment. Their patients fell into three age groups.

Group 1 included 47 children 5 years of age or younger. Four children had curves of more than 100°, 23 children had curves between 70° and 99°, and only 20 children had curves of less than 70°. Group 2 included 37 children from 6 to 10 years. Fourteen had curves of more than 100°, 13 had curves between 70° and 99°, and only ten children had curves of less than 70°. Group 3 included 23 children from 11 years of age to maturity. Twelve had curves of more than 100°, nine had curves of 70° to 99°, and only two children reached 11 years with curves of less than 70°.

From this we can conclude that without treatment virtually all progressive curves will exceed 70°, and the majority will progress to more than 100° by the age of 10 years. This severity of curvature not only produces a horrendous cosmetic deformity (Fig. 16-7D, E, and F), but also interferes with the respiratory function and results in cor pulmonale and often death in early adult life.^{25,32}

PROGNOSIS

Because of the difference in prognosis, it is important to distinguish between resolving and progressive curves at an early stage, and if necessary start immediate treatment without waiting for progression to occur. Before 1972, the only prognostic factors thought to be of value were the age of onset, the initial severity of the curve, and the presence or absence of compensatory curves. It was not, however, until 1972 when Mehta¹⁸ published her important paper on the rib-vertebra angle difference and the relationship of the rib head to the apical vertebra that it was possible for the first time to predict accurately the prognosis at an early stage when the curve was still small.

Age of Onset

Lloyd-Roberts and Pilcher¹² found that 92% of children developing a scoliosis in the first year of life had resolving curves. In Edinburgh, 83% of curves developing in the first year resolved, whereas, only 30% resolved if the curvature developed after 1 year.¹⁷ Curves developing in the first year of life therefore have a more favorable prognosis, but this is not a factor on which to base a course of treatment.

Size of Curvature

Curves greater than 35° will usually progress and very few will resolve.³ However, many curves are less than 20° when first seen and a significant number of these will also progress.

Secondary Curves

Compensatory curves are usually absent in both progressive and resolving curves when first seen at an early stage. Should compensatory curves develop, this is usually a sign that the primary curve will progress.⁴ This again is a late prognostic sign.

Rib-Vertebra Angle Difference

The rib-vertebra angle as described by Mehta is the angle formed on each side between the apical thoracic vertebra and its corresponding rib.¹⁸ The rib-vertebra angle difference is the difference between the rib-vertebral angle on the convexity of the curve subtracted from that on the concavity and may be either a positive or negative value. In a normal spine the rib-vertebra angle difference at any vertebra is zero.

To measure the rib-vertebra angle (Fig. 16-

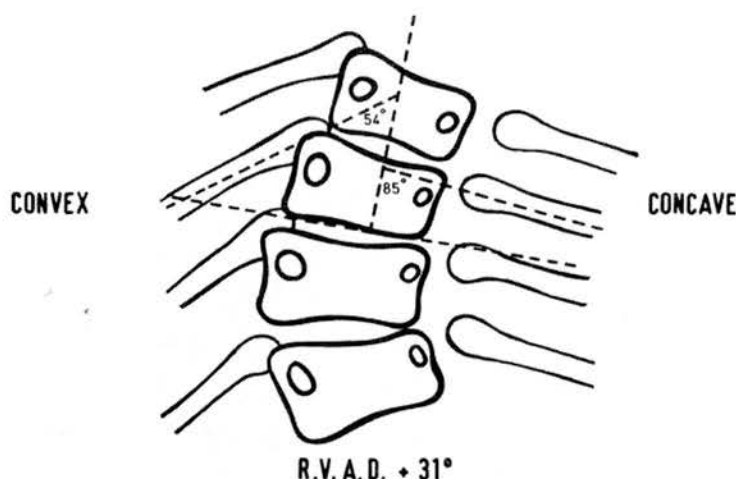


Figure 16-9. Measurement of the rib-vertebral angle difference (R.V.A.D.).

9), a good quality supine anteroposterior radiograph is required. The vertebra at the apex of the thoracic curve is selected, but if the apex of the curve is at a disk space the vertebra above is chosen. This is an important point, because significant differences can occur if the wrong vertebra is chosen. A line is drawn along the lower border of the apical vertebra and a perpendicular line is erected from its midpoint. A second line is drawn from the middle of the head of the rib associated with the apical vertebra to the midpoint of the neck of the rib just before the region of the rib where the neck expands to become the shaft. This line along the rib is the most difficult to judge, and it is important not to make the second point too lateral because this will produce the wrong inclination. The line along the rib is extended medially to intersect the perpendicular from the vertebra and subtend the rib-vertebral angle. The rib-vertebral angle on the other side of the apical vertebra is measured in the same manner.

Mehta also described the relationship between the rib head on the convexity and its vertebral body as being in either phase I or phase II.

Resolving Scoliosis

Resolving curves are nearly always thoracic and when first seen the rib-vertebral angle difference is less than 20° in 80% of the patients. After 3 months, the angle difference is reduced even though the curve itself may show a temporary increase. The usual pattern is for the rib-vertebral angle difference to decrease as the curve resolves (Fig. 16-6A and B). The rib heads on the convexity of the curve always remains separate from their vertebral bodies and this is described as phase I.

Progressive Thoracic Scoliosis

As the progressive curve deteriorates, there is an increasing downward obliquity of the ribs on the convexity, and this is greatest at the apex of the curve. This obliquity produces a small rib-vertebra angle on the convexity and as a result the angle difference is more than 20° in 80% of the patients (Fig. 16-7A). After 3 months the rib-vertebral angle difference has usually increased or remains the same. The usual pattern is for the angle difference to increase as the curve progresses. A definite diagnosis of a progressive curve can be made when the rib head on the convexity becomes overlapped by its corresponding vertebral body. This radiographic appearance, which is due to rotation of the curve, is described by Mehta as phase II and no curve however small will resolve once this sign is present.

Double Structural Scoliosis

It is common for double structural curves to present initially as a single thoracic curve without evidence of a curve or rotation in the lumbar region. The observations made by Mehta now allow us to anticipate the development of the lumbar curve before it appears. In patients who develop double structural curves, the ribs on either side of the apical vertebra in the thoracic curve are usually horizontal, arched slightly upward, and nearly symmetrical (Fig. 16-8A). As a result, the rib-vertebral angle difference is very small or may be a negative value. This may suggest a single resolving thoracic curve, but this mistake is avoided by also noting a marked obliquity or drooping of the 12th rib on the concave side of the thoracic curve. This rib is in fact on the convexity of the lumbar curve, which

is about to develop on the opposite side from the thoracic curve. Another sign of a developing lumbar curve is rotation of the lumbar vertebrae in an opposite direction to the thoracic vertebrae (Fig. 16-8B).

These observations by Mehta on the rib-vertebral angle difference and the relationship of the rib head to the vertebral body have been confirmed by Ferreira and James⁴ in an even higher percentage than she herself reported. It is now possible to recognize potentially severe curves at a very early stage and commence immediate treatment. It is also possible to reassure parents of infants with resolving curves that they are likely to disappear and so avoid unnecessary worry.

TREATMENT

An infant presenting with a possible idiopathic scoliosis must be very carefully assessed both clinically and radiographically, and the diagnosis is made by exclusion. Clinically, apart from plagiocephaly and a spinal curvature, the infant should be normal with no evidence of a muscular or neurologic disorder. Good quality spinal radiographs are essential and should be taken both with the infant supine and suspended by the arms. A postural scoliosis is excluded by noting that the curve does not fully correct on suspension. A congenital spinal deformity is excluded by carefully searching both the anteroposterior and lateral spinal radiographs for any developmental vertebral anomaly that could cause the curvature. In very young infants, a misdiagnosis is occasionally possible because a short unilateral unsegmented bar may not be apparent radiographically until it becomes fully ossified over the next few years.¹⁶ An oblique spinal radiograph that gives a true anteroposterior view of the more severely rotated spine is useful in detecting hidden congenital anomalies at the apex of the curve.

Once the diagnosis of an infantile idiopathic scoliosis is made the rib-vertebra angle difference is measured at the apex of the curve on the supine anteroposterior radiograph. By this means, it is possible to differentiate between curves that are likely to progress and those that will resolve. If the curve is less than 20° it is best to wait for another 3 months and remeasure the angle difference before making a definitive diagnosis.

Resolving curves require no treatment other than radiographic assessment at increasing intervals until the spine is straight. However, I

recommend that the child is laid prone when in its cot, and this may encourage a more speedy resolution.

The key to the successful treatment of progressive curves is to recognize their poor prognosis at an early stage and to start immediate treatment. The curve is corrected as much as possible and further deterioration is prevented by all possible conservative means until the child is considered old enough for spinal fusion. Posterior fusion is a well-established method of preventing increasing spinal deformity in adolescent scoliosis, but cannot be satisfactorily applied to very young children. It is widely accepted that posterior fusion stops all longitudinal growth in the posterior elements and may therefore cause a relative shortening of the spine as the child grows to maturity. It has also been suggested that an increasing lordosis may develop in the fused area due to continued growth of the vertebral bodies and lack of growth in the posterior elements.²³ In order to minimize these difficulties, it has been the Edinburgh policy to postpone spinal fusion until the age of 10 years unless progression of the curve cannot be controlled by other means. A long period of conservative treatment is therefore necessary to prevent deterioration of these curves, which develop before the age of 3 years and can only be satisfactorily fused after the age of 10 years. This policy is applied to all children with malignant progressive curves, but not necessarily to the few children with benign progressive curves or to double structural curves. Benign progressive curves can often be satisfactorily controlled by conservative means until later in adolescence, when a spinal fusion is more acceptable. Double structural curves affecting the thoracic and lumbar regions are usually well balanced and the cosmetic deformity is not severe. Correction and fusion of these curves results in little cosmetic improvement and the nearly total spinal fusion at an early age will produce more apparent stunting of the spine. Double curves are therefore managed conservatively until a much later age before spinal fusion, or if possible to maturity without fusion. Spinal fusion of double structural curves is only considered if the thoracic component deteriorates to more than 60° and future cardiorespiratory complications become likely.

Once the diagnosis of a progressive curve has been made, the likely prognosis and plan of treatment is discussed with the parents. The need for conservative treatment is emphasized and the parents are informed that a spinal fusion will eventually be necessary as part of the planned course of treatment.¹⁵

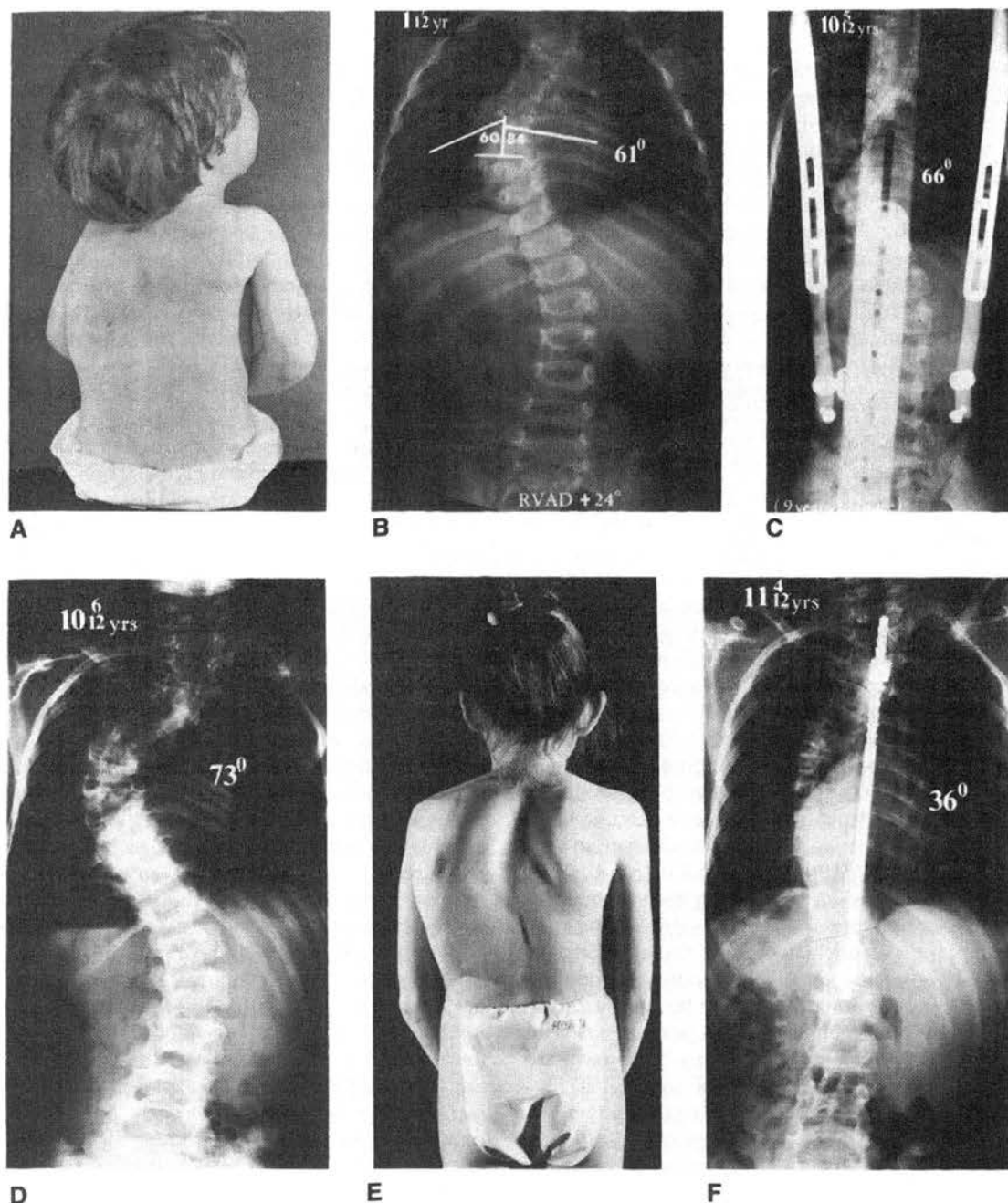
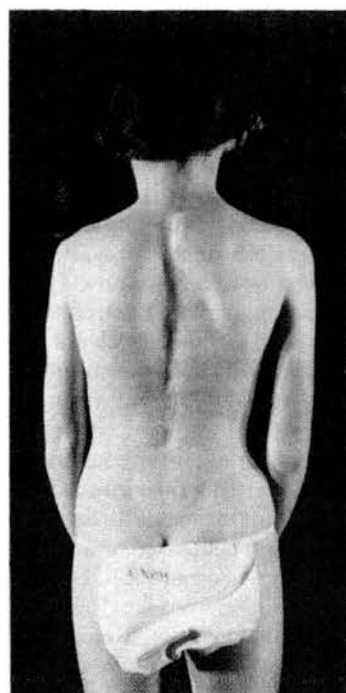
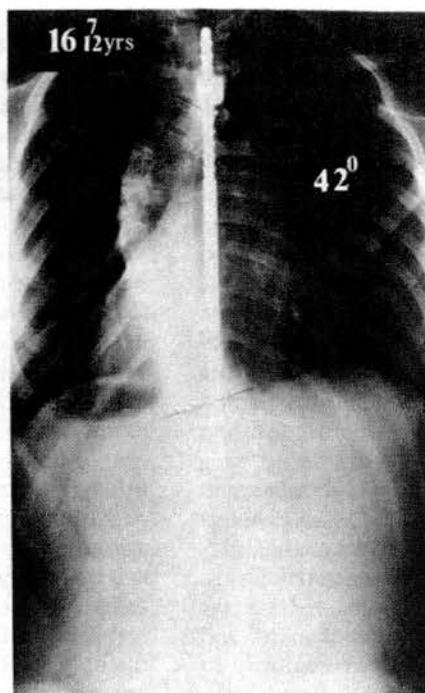


Figure 16-10. **A.** A girl, 1 year, 7 months, with a left thoracic scoliosis. **B.** The spinal radiograph shows the scoliosis to measure 61° . The rib-vertebral angle difference is $+24^{\circ}$, indicating a progressive curve. **C.** A Milwaukee brace was immediately applied and after 6 years in the brace the curve measured 49° . However, after this time, the curve became more difficult to control and by 10 years, 5 months, it measured 66° in the brace. **D and E.** Before surgery at the age of 10 years, 6 months, the curve measured 73° out of the brace, and there was a moderately severe cosmetic deformity with a large rib hump. **F and G.** Ten months after surgery there was a solid fusion and the scoliosis measured 36° . Clinically, there was a good cosmetic result and the rib hump had been reduced. **H.** At the age of 16 years, 7 months, there was a 6° loss in correction in the solid fusion. Note the narrowing of the intervertebral disk spaces due to the continued growth of the vertebral bodies anteriorly. This has caused the fused area to bulge laterally toward the convexity and pivot on the posterior fusion, giving rise to the loss of correction and increasing vertebral rotation. **I.** Clinically, there is a good cosmetic result with only mild stunting of the spine. The rib hump has, however, become slightly more prominent.



G



H



I

Corrective Plaster Jacket

Initially, all patients with progressive curves are treated in a series of corrective plaster jackets of the Risser-Cotrel type. These jackets provide the maximum amount of correction before a Milwaukee brace is fitted. It is very difficult to fit a Milwaukee brace to a little chubby baby less than 1 year old because there is no waist around which the pelvic girdle can be fitted. The plaster jacket is applied under a ketamine anesthetic with the child supine on the plaster table and longitudinal traction applied. Correction is maintained by molding the plaster beneath the occiput and mandible and over the iliac crests and rib hump. It is usually necessary to apply three plaster jackets at 6- to 8-week intervals before maximum correction is obtained. After 6 months, the plaster jacket is removed and a Milwaukee brace applied. Mehta²⁰ has found it possible to convert some benign progressive curves to resolving or stable curves by continuing with serial plaster corrections until the rib-vertebral angle difference is reduced to zero. This is not possible with malignant progressive curves, which are very much more rigid.

Brace Treatment

The Milwaukee brace has been most frequently used in the long-term conservative

management of progressive infantile idiopathic scoliosis. The type of Milwaukee brace used in young children has a padded neck ring rather than a throat mold and initially relies more on distraction as well as the action of the pad over the rib hump. The more traditional Milwaukee brace is applied when the child is older and can cooperate in an exercise program. More recently, an underarm type of Boston brace has been used with satisfactory short-term results. Because of the variation in size, both types of brace have to be individually made from a mold of the child and renewed regularly as the child grows.

A study has been made of 27 Edinburgh children who completed long-term management in a Milwaukee brace. Twenty-two children had single thoracic curves and the brace was applied at a mean age of 4 years 3 months when the mean curve was 63° (range, 29° to 105°). The mean best curvature in the brace was 36° (43% correction), and this was obtained after 1 to 2 years. After the initial correction, there was then a gradual loss of correction of approximately 1° per year. The mean rate of deterioration before the brace was applied had been 12° annually. The brace was worn for a mean period of 6 years 5 months and 70% of the children were in a brace for more than 5 years (Fig. 16-10B and C). After wearing the brace for 5 years, the rate of deterioration became greater and the final mean

curvature in the brace at the age of 10 years just before spinal fusion was 56° (11% correction). The five double curves were much less severe and the brace was not applied until a mean age of 7 years 9 months. The brace was much less effective in gaining correction in these curves, and most of the correction was lost during the adolescent growth spurt.

Electrospinal Instrumentation

Unilateral electrical stimulation of the spinal muscles on the convex side of a scoliosis, either transcutaneously or by surgically implanted electrodes, has been successful in controlling smaller curves in adolescent idiopathic scoliosis.² I have used an electrospinal orthosis to stimulate transcutaneously the muscles in infants with idiopathic scoliosis and found its success to be limited to benign progressive curves that were flexible and less than 30°. Malignant progressive curves were often more than 30° and too rigid, and none responded satisfactorily to this treatment alone. However, when the electrospinal orthosis was combined with a Milwaukee brace, it was found to be more successful. The brace was worn on a full-time basis and the electrospinal orthosis applied only at night with the electrodes over the rib hump at the site of the pad, which was removed. This combined treatment was most effective in reducing the size of the rib hump and also controlling the apparent kyphosis that was often associated with the scoliosis.

Harrington Instrumentation without Fusion

Harrington instrumentation without fusion can also be used in young children with severe progressive curves in an attempt to gain repeated corrections over a period of years before finally fusing the spine at a more suitable age. In these children, one has to accept a high incidence of instrument failure despite the additional use of a Milwaukee or Boston brace to immobilize the spine. The rod must be placed subcutaneously or within the paraspinal muscles to prevent inadvertent partial fusion of the spine on its concavity, which can occur after the usual subperiosteal exposure. In my experience this method has not been very successful because of the inability to obtain further correction after the second distraction. At this stage, the rod appears to tether the curve rather than correct it, and posterior fusion has been necessary. It is possible that the Luque technique of

segmental spinal instrumentation without spinal fusion may have a role to play in maintaining correction and allowing spinal growth in these young children until a more suitable age for fusion.

Posterior Spinal Fusion

Despite conservative treatment, all patients with malignant progressive curves eventually require a spinal fusion to control their progress. It is only with difficulty that malignant curves are controlled by conservative means until the age of 10 years, and after this they become impossible to control during the adolescent growth spurt. It has therefore been the Edinburgh policy to carry out a posterior fusion of all malignant progressive thoracic curves as a routine procedure at the age of 10 years. An even earlier fusion may be necessary if the curve cannot be controlled by other means.

The scoliosis is corrected at the time of the fusion procedure by the use of Harrington instrumentation. In severe curves it is necessary to bend the distraction rod to conform to the apparent kyphosis in the upper thoracic region. This unfortunately lessens the mechanical advantage when distraction is applied and the upper hook can easily cut out unless carefully seated.

The fusion technique is important because a previous study from our unit has shown that even a solid fusion can bend during the adolescent growth spurt, resulting in a significant loss of correction.¹⁵ In order to overcome this problem Winter²⁹ has advised supporting such young fusions in a Milwaukee brace during the period of rapid growth between the ages of 10 and 14 years. I have attempted to overcome the problem by applying an additional massive onlay of autogenous iliac bone grafts to the fusion area in an effort to produce a wide, thick fusion that would be more stable and resist bending. A prospective study was made of 24 children with idiopathic scoliosis requiring a fusion at the age of 10 years.⁷ Before operation their mean curvature was 67°, and this was reduced intraoperatively using Harrington instrumentation to a mean of 33° (51% correction). The scoliosis was then fused using a meticulous interfacetal intertransverse fusion technique²¹ followed by a deep and thorough decortication of all of the posterior spinal elements from the midline to the tips of the transverse processes. This was followed by the application throughout the fusion area of large amounts of fresh autogenous iliac bone grafts.

Postoperatively, the children were nursed free in bed for 7 to 10 days before an underarm plaster jacket was applied. They were then allowed to return to school and normal activities within 2 weeks but were not allowed to participate in contact sports. When the plaster jacket was removed after 9 to 10 months, all of the children had developed a solid fusion and there was a loss of correction of only 4° (Fig. 16-10F and G). They were then allowed unrestricted activities without support and after a mean follow-up of 4.5 years, there was a further mean loss of correction of 5° and all the spines had become stable (Fig. 16-10H and I). It is debatable whether this loss of correction is sufficiently severe to warrant the inconvenience of a further period in a brace, which would have to be worn for several years and might not be effective in stabilizing the fusion.

At final follow-up the majority of these children had passed through their adolescent growth spurt, and although there was mild to moderate stunting of the spine, only 2 of the 22 children were below the third percentile for the average height of British children of an equivalent age.^{7,26} This stunting of the spine is considered acceptable in comparison to the shortening that would have occurred if the curvature had been allowed to progress. Accurate measurements of the growth of the fusion area showed that although the posterior elements ceased to grow longitudinally, the vertebral

bodies continued to increase in height. However, the thick posterior fusion was of sufficient strength to prevent the development of a lordosis, and the increasing height of the vertebral bodies was initially accommodated by narrowing of the intervertebral disk spaces. Once these spaces had narrowed, the continued longitudinal growth caused the vertebral bodies to bulge laterally toward the convexity of the curve and also to pivot on the unyielding posterior fusion mass.⁷ This mechanism caused the postoperative loss in correction of the solidly fused curves and also an increase in the rib hump that had been partly corrected immediately following spinal instrumentation. The increasing rib hump gave the illusion of a developing thoracic lordosis, but this was not confirmed on the lateral spinal radiographs. It is possible that an anterior spinal fusion combined with the posterior fusion could prevent the vertebrae from rotating and the redevelopment of the rib hump.

CONCLUSION

The treatment of progressive infantile idiopathic scoliosis requires a long and difficult struggle to prevent increasing deformity over many years. However, with a planned course of treatment incorporating both conservative and operative means, satisfactory results can be obtained and severe deformity prevented.

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Section D

Juvenile Idiopathic Scoliosis

Juvenile Idiopathic Scoliosis

CURVE PATTERNS AND PROGNOSIS IN ONE HUNDRED AND NINE PATIENTS*

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ABSTRACT: We reviewed the medical records and radiographs of 109 consecutive patients who had juvenile idiopathic scoliosis. The sixty-seven girls and forty-two boys were a mean of six years and ten months old (range, three years and four months to nine years and eleven months old) when the curve was recognized. One hundred and four patients had a progressive curve: twenty-eight had a single mid-thoracic curve with the apex usually at the eighth thoracic vertebra (Group 1A), twenty-nine had a major mid-thoracic curve with the apex usually at the eighth thoracic vertebra and a secondary minor lumbar curve (Group 1B), twenty-seven had a single thoracic curve with the apex usually at the ninth or tenth thoracic level (Group 2), eight had a single thoracolumbar curve with the apex at the twelfth thoracic level (Group 3), and twelve had a major lumbar curve with the apex at the second or third lumbar level and a secondary minor thoracic curve (Group 4). Five patients (5 per cent) had a resolving curve.

Eighty-nine of the patients who had a progressive curve were followed to skeletal maturity. Eighty-eight patients were managed with a brace. The curve progressed at a rate of 1 to 3 degrees per year before the age of ten years and 4.5 to 11 degrees per year after the age of ten years. In sixty-seven of the eighty-four patients in Groups 1 and 2, a spinal arthrodesis was performed before the age of fifteen years (mean age, eleven years and ten months; range, nine years and three months to fourteen years and eight months), at which time the mean curve was 47 degrees (range, 24 to 90 degrees). Eight patients were not seen by us until they were fifteen years old or more, at which time the thoracic curve was 74 to 120 degrees; a spinal arthrodesis was done in seven. The curves in Groups 3 and 4 had a more benign prognosis, and only three patients in these two groups had an arthrodesis.

The pattern of the final curve was not always apparent at an early stage, and there was extension of the primary curve or development of secondary structural curves with progression. The chief prognostic

feature at an early stage was the level of the most rotated vertebra at the apex of the primary curve, and the final pattern of deformity was defined by the level of the caudad neutral vertebra in the primary thoracic curve.

James classified idiopathic scoliosis on the basis of the patient's age when the scoliosis was first identified: it was classified as infantile at an age of less than three years, as juvenile at an age of three to ten years, and as adolescent at an age of ten years to skeletal maturity. In contrast to the situation with infantile and adolescent idiopathic scoliosis¹⁶, relatively little is known about the curve patterns and the natural history of juvenile idiopathic scoliosis. Tolo and Gillespie used a brace on forty-two of fifty-nine patients who had juvenile idiopathic scoliosis and performed a spinal arthrodesis on the other seventeen. Mannherz et al. followed forty-three patients who had juvenile idiopathic scoliosis to maturity. The curve progressed in twenty-one of the thirty-one patients who had been managed with a brace; fourteen of the thirty-one needed operative management at a mean age of fifteen years for a mean curve of 51 degrees (range, 33 to 87 degrees). Figueiredo and James reported that thirty-one of ninety-eight patients needed an arthrodesis of the spine and that seventy-six of the ninety-eight patients had a right-sided curve. We evaluated the different curve patterns and their course in 109 patients who had juvenile idiopathic scoliosis. Our purpose was to identify any characteristics that could predict the course of the curve.

Materials and Methods

We reviewed the medical records and radiographs of 109 consecutive patients with juvenile idiopathic scoliosis who had been followed between 1970 and 1992 at the Princess Margaret Rose Orthopaedic Hospital, Edinburgh. The study cohort was derived from 1362 patients who had been evaluated for idiopathic scoliosis during this time-period. Ninety-one of the more recently seen patients have been under the care of the senior one of us (M. J. McM.). During this period, the mean prevalence of new patients who had juvenile idiopathic scoliosis was 8 per cent (range, 4 to 22 per cent) per year.

The patients were asked to return for follow-up every four to six months, depending on the severity of the

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TABLE I
CLINICAL DATA ON THE ONE HUNDRED AND NINE PATIENTS WHO HAD JUVENILE IDIOPATHIC SCOLIOSIS*

	Progressive Curves					Resolving Curves
	Group 1A	Group 1B	Group 2	Group 3	Group 4	
No. of patients	28	29	27	8	12	5
Mean age at present. (yrs. + mos.)	7 + 3	6 + 9	7 + 3	5 + 11	7 + 10	4 + 2
Ratio of girls to boys	13:15	19:10	17:10	6:2	11:1	1:4
Thoracic curves						
Side (right:left)	17:11	24:5	18:9	5:3	11:1	3:2
Cobb angle (degrees)†	39 (10-49)	39 (17-68)	40 (19-56)	28 (18-45)	22 (10-57)	23 (15-40)
Rotation of apical vertebra (degrees)†	24 (15-30)	22 (12-45)	22 (15-45)	16 (5-20)	10 (5-15)	15 (10-30)
Rib-vertebra angle difference (degrees)†	34 (16-40)	35 (15-42)	38 (16-45)	21 (12-29)	10 (7-19)	9 (5-20)
Kyphosis (degrees)†	19 (12-20)	18 (15-20)	19 (15-23)	19 (13-20)	29 (23-35)	23 (15-29)
Lumbar curves						
Side (right:left)	NA	5:24	NA	NA	1:11	NA
Cobb angle (degrees)†	NA	24 (10-45)	NA	NA	30 (18-58)	NA
Rotation of apical vertebra (degrees)†	8 (0-10)	17 (5-20)	NA	NA	17 (11-20)	NA
Lordosis (degrees)†	43 (33-49)	47 (32-50)	45 (30-50)	45 (39-48)	50 (45-60)	43 (20-50)

*NA = not applicable.

†The values are given as the mean, with the range in parentheses.

curve. Anteroposterior and lateral radiographs of the spine were made, with the patient standing, at the initial visit and at each follow-up visit. Side-bending radiographs of the spine were not made routinely. Initial measurements were made on all of the radiographs by the senior one of us, and then measurements were made by the other one of us (C. M. R.). The final values were based on a consensus between us; intraobserver and interobserver errors were not estimated.

The levels of the neutral, end, apical, and stable⁷ vertebrae were recorded, the scoliotic curve was measured², and the rib-vertebra angle difference¹⁰ and the vertebral rotation¹² were measured at the apical vertebra of the thoracic curve (Table I). Spinal decompensation, or list, was measured on the anteroposterior radiograph as the amount of lateral displacement of the seventh cervical vertebra with respect to the central sacral line. Lateral translation of the thoracic cage, distinct from list, was the degree of displacement of the thorax from the central sacral line¹⁴. Sagittal alignment was assessed on the lateral radiograph by measurement of the thoracic kyphosis from the third to the twelfth thoracic vertebra and the lumbar lordosis from the first to the fifth lumbar vertebra¹.

Results

Age, Gender, and Side of the Curve

The mean age at the time of the diagnosis of the curve in the 109 patients was six years and ten months (range, three years and six months to nine years and eleven months). There were sixty-seven girls and forty-two boys (Table I). The mean age of the boys when they were initially seen was five years and eight months (range, three years and six months to nine years and seven months), which was slightly younger than the mean age of the girls (seven years and two months; range, three years and eight months to nine years and eleven months) when they were first seen. The ratio of

girls to boys was 1:1.6 in the age-group of less than six years old and 2.7:1 in the age-group of more than six years old. Female predominance (11:1) was most pronounced in the group that had a primary progressive lumbar curve.

The primary progressive thoracic and thoracolumbar curves and the resolving curves were right-sided in sixty-seven children, and left-sided in thirty (Table I). Eleven of the twelve primary progressive lumbar curves were left-sided. In the group of children who were less than six years old, there were an equal number of right and left-sided curves, whereas the older children had more right-sided curves (ratio, 3.9:1). There was no meaningful difference in the mean age at the time of diagnosis among the four groups of children who had a progressive curve; however, the children who had a resolving curve were younger when they were first seen (mean age, four years and two months; range, three years and six months to five years and six months) than those who had a progressive curve.

Pattern of Progressive Curves

Attempts to classify the curves with use of the radiographs that were made at the time of the initial visit were unsuccessful, as the deformity often changed as a result of extension of the primary curve or the development of secondary curves that later became fixed. The apical vertebra remained constant, even when the curve extended to involve additional vertebrae, and it was used to predict the final pattern of the deformity. The development of secondary curves and the degree of spinal imbalance were similar for patients in whom the apical vertebra was at the same level; hence, these patients were grouped together (Figs. 1-A and 1-B).

Group 1

Fifty-seven of the children had a progressive thoracic curve with the apex usually at the eighth thoracic

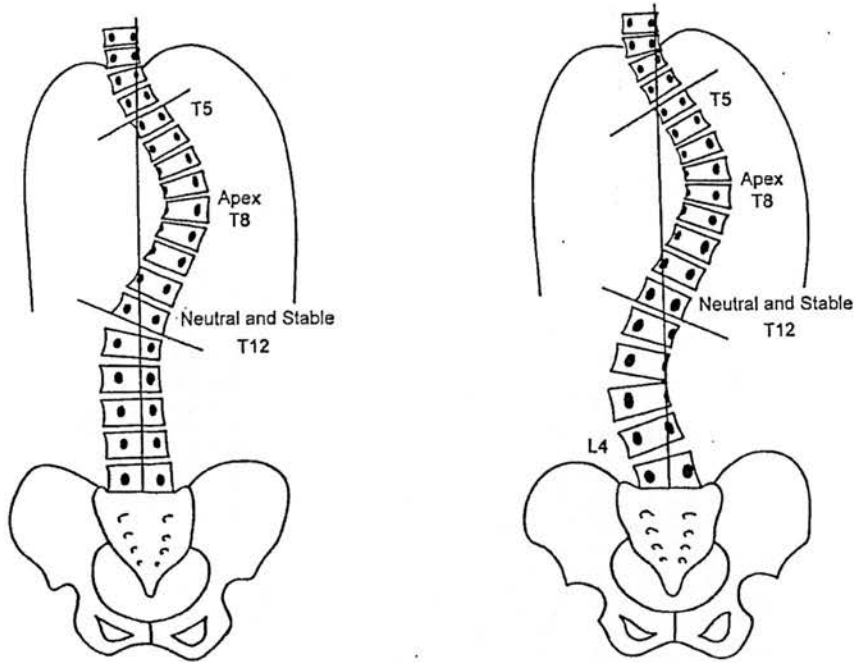


FIG. 1-A

Figs. 1-A and 1-B: Line diagrams of the different progressive curve patterns in juvenile idiopathic scoliosis. Fig. 1-A: Groups 1A (left) and 1B (right).

level (Group 1). As the curve progressed, the caudal neutral vertebra remained at the eleventh or twelfth thoracic level and the lumbar vertebrae rotated in a direction opposite to the rotation of the thoracic curve. There were two curve patterns: a single mid-thoracic curve (Group 1A) and a major mid-thoracic curve with

a secondary minor lumbar curve (Group 1B).

There were twenty-eight patients in Group 1A. The apical vertebra was at the eighth thoracic level in twenty-four patients and at the ninth thoracic level in four. The curves extended from a neutral vertebra at the fourth or fifth thoracic level to a neutral vertebra

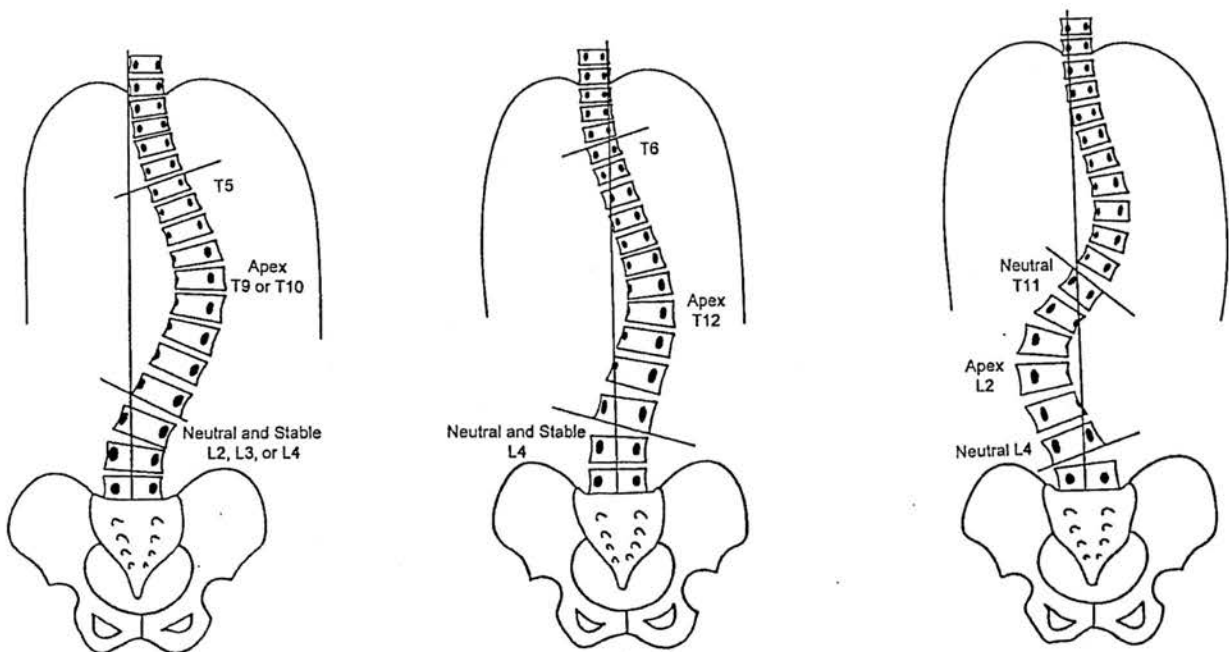


FIG. 1-B

Groups 2 (left), 3 (middle), and 4 (right).

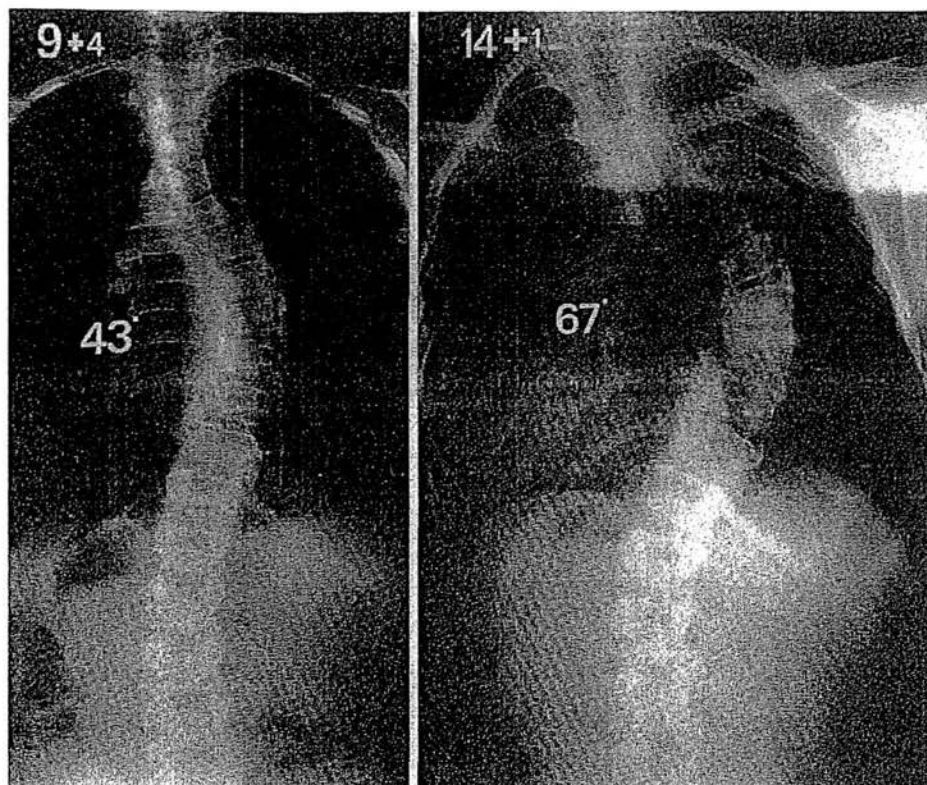


FIG. 2-A

FIG. 2-B

Figs. 2-A and 2-B: Radiographs showing progression of a Group-1A curve.

Fig. 2-A: The boy was nine years and four months old when he was first seen. The right-sided thoracic curve was 43 degrees, and the apex was at the eighth thoracic level.

Fig. 2-B: When the boy was fourteen years and one month old, the curve had progressed to 67 degrees. The caudad neutral vertebra remained at the twelfth thoracic level.

at the eleventh or twelfth thoracic level, and the stable vertebra was usually at the twelfth thoracic or first lumbar level. As the thoracic curve progressed, a slight secondary lumbar curve developed, but no patient had a lumbar curve that crossed the mid-sacral line. The body of the fourth lumbar vertebra remained relatively square to the sacrum (Figs. 2-A and 2-B). Lateral translation of the thoracic cage increased as the curve progressed because of the absence of a compensatory lumbar curve, and there was a list toward the convexity of the curve. There was hypokyphosis of the thoracic spine and normal lumbar lordosis.

Twenty-nine patients were in Group 1B. The apical vertebra was at the eighth thoracic level in twenty-six patients and at the seventh thoracic level in three. In the early stage, the thoracic curves appeared similar to those in Group 1A, with the caudad neutral and stable vertebrae at either the eleventh or the twelfth thoracic level. However, as the thoracic curve progressed, a lumbar curve developed, with the apex at the second or third lumbar level and the neutral vertebra at the fourth lumbar level, which was always tilted toward the convexity of the curve (Figs. 3-A and 3-B). The two curves ultimately progressed at the same rate, and spinal balance was restored; this resulted in a less severe

cosmetic deformity than in Group 1A; however, the sagittal profile was the same in all of the curves in Group 1.

Group 2

Group 2 consisted of twenty-seven patients who had a single progressive thoracic curve with the apex at the ninth thoracic vertebra (twelve patients), the tenth thoracic vertebra (eleven patients), or the eighth thoracic vertebra (four patients) (Fig. 1-B). In this group, unlike in Group 1, the lumbar vertebrae rotated into the primary curve with progression, resulting in a shift of the caudad neutral vertebra to a more caudad level (the second lumbar level in ten patients, the third lumbar level in three, and the fourth lumbar level in fourteen). The stable vertebra was at the second lumbar level in eight patients, the third lumbar level in four, and the fourth lumbar level in fifteen. A compensatory lumbar curve did not develop in these patients (Figs. 4-A and 4-B), which resulted in a severe list, elevation of the ipsilateral shoulder, translation of the thoracic cage to the side of the curvature, and prominence of the contralateral hip. The more caudad the neutral vertebra in the primary curve, the more severe the deformity. The sagittal profile of the tho-

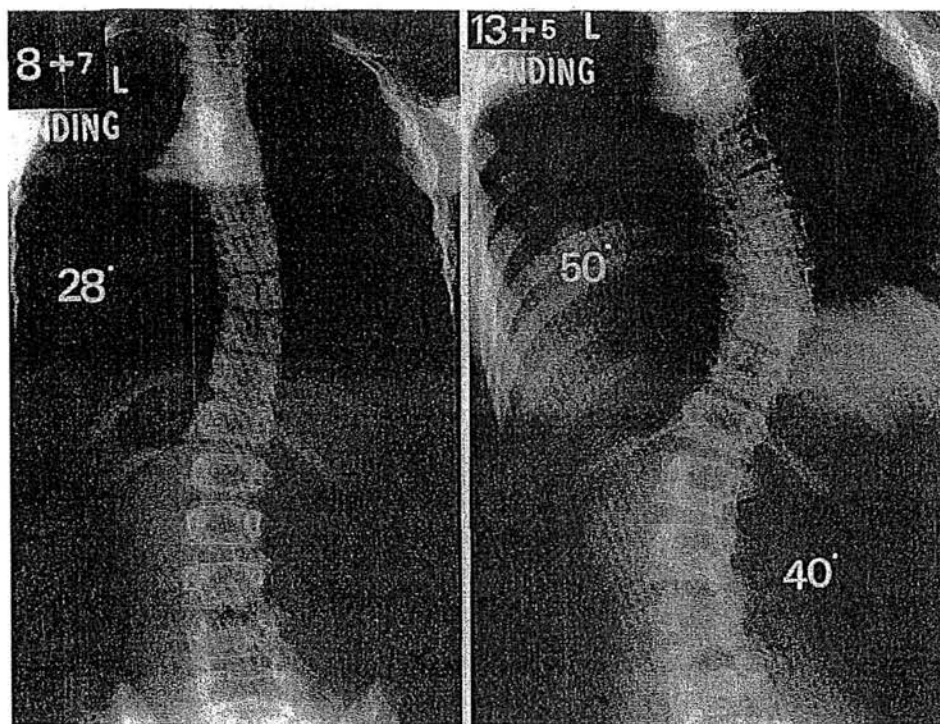


FIG. 3-A

FIG. 3-B

Figs. 3-A and 3-B: Radiographs showing progression of a Group-1B curve.

Fig. 3-A: The girl was eight years and seven months old when she was first seen. The right-sided thoracic curve measured 28 degrees, and the apex was at the eighth thoracic level.

Fig. 3-B: When the girl was thirteen years and five months old, the thoracic curve had increased to 50 degrees and there was a secondary lumbar curve measuring 40 degrees. The stable vertebra and caudad neutral vertebra of the thoracic curve remained unchanged at the twelfth thoracic level.

racic spine was hypokyphotic, and lumbar lordosis was normal.

Group 3

Group 3 included eight patients who had a long, tapering progressive thoracolumbar curve. The apex was at the twelfth thoracic level, and the neutral vertebrae were at the fifth or sixth thoracic level and at the fourth lumbar level (Fig. 1-B). The neutral vertebra at the fourth lumbar level was always tilted into the curve. The translation of the thoracic cage and the list were not as severe as in Group 2. There was hypokyphosis of the thoracic spine, and lumbar lordosis was normal.

Group 4

Group 4 consisted of twelve patients who had a primary progressive lumbar curve and a secondary minor thoracic curve. The apex was at the second lumbar level in ten patients and at the third lumbar level in two (Fig. 1-B). In the early stage, the lumbar curve extended from the eleventh thoracic to the fourth lumbar vertebra with slight, if any, deformity in the thoracic region. As the lumbar curve progressed, a compensatory thoracic curve developed. The eleventh or twelfth thoracic vertebra remained the neutral vertebra between these two curves. As the thoracic curve developed, spinal alignment was maintained and the cosmetic deformity

was slight. These patients had normal thoracic kyphosis and increased lumbar lordosis.

Size and Progression of the Curve and Treatment

Patients who were seen before the age of ten years were usually observed without treatment for six to nine months. A brace was used if the curve was more than 30 degrees. A Milwaukee brace was used until 1976; after that time, a modification of the Boston spinal orthosis was employed. The best correction of the curve in the brace usually occurred within the first six months, and the greatest degree of correction was seen in children who were less than six years old. Initially, the brace was worn full-time; however, if the curve was less than 30 degrees after the brace had been worn for six months to one year, the patient was allowed to take the brace off for four hours each day for four months. If the curve progressed, the brace was again worn full-time. Operative intervention was recommended if the child was more than ten years old and had a curve of more than 40 degrees that could not be controlled in the brace.

Group 1A

In Group 1A (twenty-eight patients), the diagnosis was made at a mean age of seven years and three months (range, three years and eight months to nine years and eight months). Initially, the mean curve was

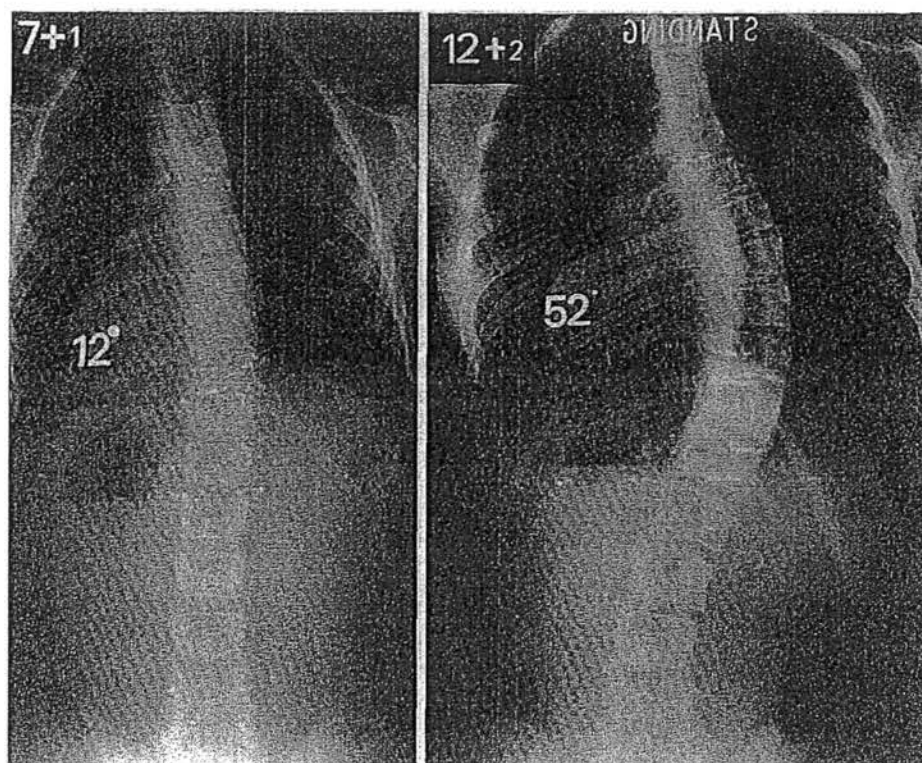


FIG. 4-A

FIG. 4-B

Figs. 4-A and 4-B: Radiographs showing progression of a Group-2 curve.

Fig. 4-A: The girl was seven years and one month old when she was first seen. The right-sided thoracic curve measured 12 degrees, and the apex was at the ninth thoracic level.

Fig. 4-B: When the child was twelve years and two months old, the curve had progressed to 52 degrees and additional lumbar vertebrae had been incorporated into the thoracic curve, resulting in a caudal shift of the caudal neutral vertebra to the fourth lumbar level.

39 degrees (range, 10 to 49 degrees). Twenty-one patients were managed with a brace for a mean of four years and three months (range, nine months to six years and six months); the curve initially decreased but subsequently progressed at a mean rate of 2 degrees per year before the age of ten years. The curve progressed more rapidly (mean rate, 8 degrees per year; range, 6 to 15 degrees per year) after the age of ten years. All twenty-one patients had an arthrodesis of the spine at a mean age of eleven years and nine months (range, nine years and three months to fourteen years and eight months); at that time, the mean thoracic curve was 46 degrees (range, 28 to 82 degrees). Four patients were first seen by us at a mean age of seventeen years and six months (range, sixteen years and six months to twenty years), at which time the curves were 74, 78, 80, and 120 degrees; an arthrodesis of the spine was done in each patient. Three patients, eight years and three months to ten years old, were still being managed with a brace at the time of writing.

Group 1B

In Group 1B (twenty-nine patients), the diagnosis was made at a mean age of six years and nine months (range, three years and ten months to nine years and eleven months). Initially, the mean thoracic curve was 39

degrees (range, 17 to 68 degrees) and the mean lumbar curve was 24 degrees (range, 10 to 45 degrees). Twenty-five patients were managed with a brace for a mean of five years and six months (range, two to ten years). Before the age of ten years, there was an initial decrease of both curves, but then progression occurred at a mean rate of 1.5 degrees per year for the thoracic curves and 1 degree per year for the lumbar curves. After the age of ten years, there was much more rapid progression of the curve: a mean rate of 7.5 degrees per year for the thoracic curves and 6.5 degrees per year for the lumbar curves.

Twenty-one patients had an arthrodesis of the spine at a mean age of twelve years and six months (range, nine years and eleven months to fourteen years and six months); at that time, the mean thoracic curve was 48 degrees (range, 24 to 86 degrees) and the mean lumbar curve was 34 degrees (range, 16 to 50 degrees). Four patients were first seen by us when they were fifteen years to sixteen years and eight months old; the thoracic curves were 78, 85, 86, and 95 degrees, and the lumbar curves were 46, 52, 62, and 73 degrees. Three of these patients had an arthrodesis of the spine, and the fourth declined additional treatment. Only one patient in this group reached skeletal maturity without rapid progression of the curve during the adolescent growth spurt. Three patients, nine years and three months to

eleven years old, were still wearing a brace at the time of writing.

Group 2

In Group 2 (twenty-seven patients), the diagnosis was made at a mean age of seven years and three months (range, three years and nine months to nine years and six months). The mean thoracic curve was 40 degrees (range, 19 to 56 degrees). Twenty-six children were managed with a brace for a mean of four years and three months (range, nine months to nine years and three months). After initial partial correction in the brace, the curves progressed at a mean rate of 3 degrees per year before the age of ten years and 11 degrees per year after the age of ten years. Twenty-five of the twenty-six patients had an arthrodesis of the spine, at a mean age of eleven years and four months (range, nine years and three months to thirteen years and eleven months); at that time, the mean thoracic curve was 47 degrees (range, 29 to 90 degrees). The remaining patient, who was twelve years old and had a 35-degree curve when first seen, was still wearing a brace at the time of writing. The twenty-seventh patient was seven years old at the time of the initial evaluation, and had a 34-degree curve that was not treated. At the time of the most recent follow-up examination, she was thirteen years and four months old and the curve was 53 degrees. The family declined all forms of treatment, despite a marked rib hump, spinal decompensation, and lateral translation of the thoracic cage.

Group 3

In Group 3 (eight patients), the patients were first seen at a mean age of five years and eleven months (range, three years and four months to nine years). The mean thoracolumbar curve measured 28 degrees (range, 18 to 45 degrees). One patient, who was seven years old at the time of the initial examination, had a 20-degree curve that was not treated. At skeletal maturity, the curve was 31 degrees and there was a mild cosmetic deformity. Two children, who initially had curves of 18 and 23 degrees, reached skeletal maturity without needing a spinal arthrodesis. One wore a brace for five years and six months, and the curve was 17 degrees at the age of sixteen years. The other wore a brace for nine years and one month and had a curve of 33 degrees at the age of seventeen years and three months. Three patients had worn a brace for a mean of five years and were still wearing a brace at the time of writing; the curve had decreased from 37 to 17 degrees by the age of twelve years in one child, from 45 to 19 degrees by the age of eight years in another, and from 40 to 14 degrees by the age of nine years in the third. Two children had a spinal arthrodesis after progression of the curve in the brace. Both children were nine years old when they were first seen; the curve progressed from 22 to 33 degrees by the age of fourteen years in one and from 28 to 36 degrees

by the age of twelve years and seven months in the other. In retrospect, both operations may have been done prematurely.

Group 4

In Group 4 (twelve patients), the diagnosis was made at a mean age of seven years and ten months (range, five years to nine years and three months). The mean thoracic curve was 22 degrees (range, 10 to 57 degrees), and the mean lumbar curve was 30 degrees (range, 18 to 58 degrees). Only one patient had a spinal arthrodesis. At the time of the initial examination, this child was nine years old, the lumbar curve was 58 degrees, and the thoracic curve was 57 degrees. The arthrodesis was done at the age of eleven years and three months, at which time the lumbar curve was 86 degrees and the thoracic curve was 80 degrees.

The remaining eleven patients were managed with a brace for a mean of seven years and eight months (range, one year and six months to ten years and two months). After initial partial correction in the brace, the mean rate of progression was 1.5 degrees per year for the lumbar curves and 2 degrees per year for the thoracic curves before the age of ten years; after this time, the curves progressed more rapidly (5 and 4.5 degrees per year, respectively). Weaning from the brace commenced at a mean age of thirteen years and five months (range, eleven years and one month to fourteen years and six months), at which time the mean lumbar curve was 36 degrees (range, 18 to 80 degrees) and the mean thoracic curve was 39 degrees (range, 26 to 86 degrees). Seven of these patients were seen at skeletal maturity, at which time the mean lumbar curve was 35 degrees (range, 28 to 42 degrees) and the mean thoracic curve was 31 degrees (range, 25 to 40 degrees). The spine was well balanced in all of these patients, and operative treatment was not necessary.

Resolving Curves

In the five patients who had a resolving curve, the diagnosis was made at a mean age of four years and two months (range, three years and six months to five years and six months). The mean curve was 23 degrees (range, 15 to 40 degrees), with slight vertebral rotation. Three of the five patients had a thoracic curve, and two had a thoracolumbar curve. The patients were managed with a brace, and the curves decreased to less than 10 degrees at a mean of four years and six months (range, two years to six years and eight months). The curves had remained stable at the time of the latest follow-up evaluation, at skeletal maturity.

Rib-Vertebra Angle Difference and Rotation of the Apical Vertebra

The mean rib-vertebra angle difference in the patients who had a progressive thoracic or thoracolumbar curve was 31 degrees (range, 12 to 45 degrees). In the

patients who had a resolving curve, the mean difference was 9 degrees (range, 5 to 20 degrees) and the difference decreased with time. In Groups 1A, 1B, and 2, both measurements changed only slightly before the age of ten years; however, they appeared to increase in proportion to the progression of the curve during the adolescent growth spurt. In Group 3, the changes were smaller and more gradual, regardless of age.

Discussion

At our institution, the prevalence of patients with juvenile idiopathic scoliosis has remained relatively constant (mean, 8 per cent per year) as a proportion of patients with idiopathic scoliosis (infantile, juvenile, and adolescent). In his London clinic, James found a prevalence of juvenile scoliosis of 12 per cent (sixteen of 134). In the United States, where infantile scoliosis is relatively uncommon¹⁵, the prevalence of juvenile scoliosis has been reported to be 13 to 16 per cent (forty-four [13 per cent] of 335¹³, twenty-six [15 per cent] of 169¹¹, and twenty [16 per cent] of 123⁶). In our study, the patients were first seen at a mean age of six years and ten months, and the over-all ratio of girls to boys was 1.6:1. Others have reported a similar age at onset, with a higher proportion of girls: the ratio of girls to boys has ranged from 3.8:1 in one study⁹ to 4.4:1 in another study¹⁷. We identified five types of curves that could be classified only as they progressed.

Juvenile scoliosis develops before a child is ten years old and progresses slowly during the period of steady spinal growth. However, after the age of ten years, progression is rapid during the period of accelerated spinal growth, despite the use of a brace. In our study, the curve progressed in 104 (95 per cent) of the 109 patients. Seventy-seven of the eighty-nine patients who were followed until skeletal maturity had an arthrodesis of the spine to prevent additional progression.

In the early stage, the level of the most rotated vertebra at the apex of the primary curve was most closely associated with the prognosis. The level of the caudad neutral vertebra in the thoracic curves could help to predict the final deformity. It has been suggested that factors that are associated with a poor prognosis (progression of the curve) in juvenile idiopathic scoliosis include a serial increase in the rib-vertebra angle difference at the apex of the primary curve, a thoracic kyphosis of less than 20 degrees, and a left-sided curve in a boy^{4,9,17}. We did not find any of these factors to be of value. In our patients, all of the primary thoracic curves were hypokyphotic and the rib-vertebra angle difference increased as the curve progressed, reflecting increasing vertebral rotation at the apex of the curve.

Initially, the curves in Group 1 appeared to be very similar, but two different types of deformity developed with progression of the curve. Nonetheless, the apical vertebra usually remained at the eighth thoracic level and the neutral vertebra remained at the eleventh or

twelfth thoracic level. We do not know why only a single structural curve developed in some of these patients (Group 1A) while a double structural curve developed in others (Group 1B). This was not related to management with a brace (because the double-curve pattern was present before the brace was used) or to the radiographic characteristics of the curve (the severity of the vertebral rotation in the primary thoracic curve, the rib-vertebra angle difference, or the sagittal deformity). Both types of thoracic curves had a poor prognosis, and most could not be controlled adequately in a brace. All twenty-five Group-1A patients who had reached skeletal maturity and twenty-four of the twenty-six Group-1B patients who had reached skeletal maturity had operative management.

The curves in Group 2 differed from the curves in Group 1 because, with progression, additional lumbar vertebrae rotated into the primary thoracic curve. This resulted in a shift of the caudad neutral and stable vertebrae to a more caudad portion of the lumbar spine, leaving insufficient room for the development of a compensatory lumbar scoliosis. The patients in Group 2 had the most pronounced cosmetic deformity because of greater lateral translation of the thoracic cage and the tendency for a list to develop toward the convexity of the thoracic curve. The more caudad the caudad neutral vertebra, the more severe the deformity. The curves in which the fourth lumbar vertebra was the last vertebra tilted into the primary curve were similar to a type-IV adolescent curve, according to the classification of King et al.; however, curves in which either the second or the third lumbar vertebra was the last vertebra to be tilted into the primary curve were intermediate between types III and IV curves according to King et al. Operative management was necessary for all of the twenty-five patients in Group 2 who had reached skeletal maturity.

Group-3 curves progressed slowly, and there was only a mild list and slight lateral translation of the thoracic cage. The curves were very mobile and were easily controlled in a brace; three patients had a curve of less than 35 degrees when they reached skeletal maturity. A spinal arthrodesis was performed to prevent progression of the curve in only two patients, who had curves of 33 and 36 degrees; in retrospect, these operations may not have been necessary.

In Group 4, the apical vertebra was at the second or third lumbar level and the curve initially presented as primary lumbar scoliosis. However, as the lumbar curve progressed, a second structural curve developed on the opposite side in the thoracic region, producing a double-curve pattern and thereby maintaining spinal balance. This pattern of deformity, which was relatively uncommon, occurred in twelve patients and was similar to a type-I adolescent curve according to the classification of King et al. This type of curve had a relatively benign prognosis, and only one patient, who had not

been managed with a brace, needed a spinal arthrodesis. The remaining patients had a well balanced spine with a relatively mild cosmetic deformity when they were seen at the most recent follow-up evaluation.

The single lumbar curves seen in adolescent idiopathic scoliosis were not found in our patients who had juvenile idiopathic scoliosis. However, the prevalence of single lumbar curves in other studies of juvenile scoliosis has been reported to be 12 per cent (seven of fifty-nine patients¹⁷ and five of forty-three patients⁹). It is possible that many of these were actually Group-4 curves and the relevance of the thoracic curve was not appreciated.

Five patients (5 per cent) had a curve that decreased or resolved; this compares with rates ranging from 52 per cent (sixty-eight of 132)³ to 92 per cent (ninety-two of 100)⁸ that have been associated with infantile idiopathic scoliosis. In our series, the resolving juvenile curves, which were seen before the children were six years old, were similar to resolving infantile curves in that they averaged 23 degrees, there was little vertebral rotation, and the rib-vertebra angle difference was less than 20 degrees¹⁰. The five curves were treated with a

brace, and they decreased to less than 10 degrees at a mean of four years and six months and remained stable throughout the adolescent growth spurt.

In most of our patients, the final curve was similar to adolescent idiopathic scoliosis. A few of our patients may have had an infantile idiopathic curve that was not recognized before the child was three years old. The etiologies of juvenile and adolescent idiopathic scoliosis may be similar, but juvenile scoliosis develops before the age of ten years. There is a rapid increase in the deformity during the adolescent growth spurt, and most patients need operative management early in the teenage years. In contrast, adolescent idiopathic scoliosis appears to develop from a straight spine during the adolescent growth spurt and is therefore subjected to a shorter period of accelerated growth of the spine before it stabilizes at skeletal maturity; only a few of these patients need operative management. We believe that the difference in prognosis warrants consideration of juvenile idiopathic scoliosis as a separate and distinct entity from adolescent idiopathic scoliosis.

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THE EFFECT OF THE ADOLESCENT GROWTH SPURT ON EARLY POSTERIOR SPINAL FUSION IN INFANTILE AND JUVENILE IDIOPATHIC SCOLIOSIS

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Twenty-four children with infantile or juvenile idiopathic scoliosis had their spines corrected and solidly fused posteriorly before the age of eleven years. The growth of the fusion area was then accurately measured for a mean of 4.5 years during the adolescent growth spurt. During this period all longitudinal growth in the posterior elements ceased. The vertebral bodies continued to grow anteriorly, but the thick posterior fusion prevented the development of a lordosis. Initially the anterior growth was accommodated by narrowing of the intervertebral disc spaces, but eventually the vertebral bodies bulged laterally towards the convexity and pivoted on the posterior fusion, giving rise to loss of correction, increasing vertebral rotation and recurrence of the rib hump.

Posterior spinal fusion with Harrington instrumentation is a widely accepted method of correcting and preventing increasing deformity in growing patients with scoliosis. Risser *et al.* (1966) stated that due to the biologic plasticity of bone, the posterior fusion was elongated by the tension stress of continued growth of the unfused vertebral bodies anteriorly. It is, however, more widely accepted that posterior fusion stops all longitudinal growth in the posterior elements and may therefore cause a relative shortening of the spine as the child grows to maturity (Winter 1971). Ponseti and Friedman (1950) also suggested that an increasing lordosis may develop in the fused area due to the continued growth of the vertebral bodies and the lack of growth in the posterior elements. Tanner, Whitehouse and Takaishi (1966) showed that the longitudinal rate of growth of the spine is not constant: there is a period of accelerated growth which occurs in British girls between the ages of 10.5 and 13.5 years, and in boys between 12.5 and 15.5 years. Little is known of the effect of this adolescent growth spurt on the spines of children which have been solidly fused posteriorly before its onset. Progressive infantile and juvenile idiopathic curves commence before the age of 10 years and may require an early posterior fusion to control their progression. In the few previous studies of

early posterior spinal fusion for scoliosis the large majority of patients had congenital scoliosis, and there were relatively few with progressive infantile or juvenile idiopathic curves (Moe, Sundberg and Gustilo 1964; Winter 1971; Winter and Moe 1982; Letts and Bobechko 1974). In congenital scoliosis the longitudinal growth of the spine is already impaired both anteriorly and posteriorly by the congenital unilateral failure of formation or segmentation of the vertebrae. In idiopathic scoliosis vertebral growth is relatively unimpaired and the effect of posterior fusion before the adolescent growth spurt may be very different.

Posterior spinal fusion had been recommended at the age of 10 years in children with progressive infantile and juvenile idiopathic scoliosis, to control the more rapid deterioration which usually occurs during the adolescent growth spurt and cannot be adequately controlled by bracing (James 1967). A previous study by one of us of 12 children with progressive infantile idiopathic curves which were solidly fused at ten years of age showed that after a mean of seven years they had lost a mean of 17 degrees of correction before becoming stable (McMaster and Macnicol 1979). Nearly all of this loss occurred in the first few years, and was due to bending of the solid but plastic fusion mass during the adolescent growth spurt. Harrington instrumentation was not used in these children and was seldom used in the few other reports of early posterior spinal fusion for idiopathic scoliosis which also recorded a relatively high loss of correction (Moe *et al.* 1964; Winter 1971; Letts and Bobechko 1974).

This paper is concerned with early posterior fusion of progressive infantile and juvenile idiopathic scoliosis, and its purpose is twofold. Firstly to report a prospective study by the senior author of the effect of Harrington

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instrumentation and a massive onlay of autogenous iliac bone grafts in an attempt to produce a strong thick posterior fusion which would resist bending during the adolescent growth spurt. Secondly to detect any adverse effects such as relative stunting of growth, increasing lordosis or rotational deformities of the spine which might result from early posterior fusion in these patients.

CLINICAL MATERIAL

At the Princess Margaret Rose Orthopaedic Hospital, Edinburgh, during the period May 1975 to April 1980, 24 patients with infantile or juvenile idiopathic scoliosis underwent posterior spinal fusion with Harrington instrumentation before the age of 11 years. All of these operations were performed by one of us (M. McM), using the same technique of correction and fusion. Twenty-three patients were recalled and reviewed personally by both authors after an average of 4.5 years (range 2 years to 7 years and 2 months) after operation. Of the 23 patients reviewed, 15 had progressive infantile idiopathic scoliosis (age of onset between 3 months and 3 years) and eight had juvenile idiopathic scoliosis (age of onset between 8 years and 9 years 6 months). There were 14 boys and nine girls. The mean age at first presentation to the scoliosis clinic was 5.5 years (range one year and 6 months to 10 years and 6 months), and all except two patients were initially treated in a Milwaukee brace for an average of 4.5 years (range one to 8 years) before operation. The mean age at operation was 10 years and one month (range 8 years and 6 months to 11 years). Twenty-two patients had single thoracic curves; one had a thoraco-lumbar curve. Thirteen curves were to the left and 10 to the right.

METHODS

Operative technique

The scoliosis was corrected by the Harrington distraction system with the hook inserted into the neutral vertebra at either end of the curve. A single distraction rod was used without the compression system (Harrington 1962). There was no pre-operative traction nor the use of corrective plaster jackets. The spine was fused posteriorly from the upper to the lower Harrington hooks by the interfacetal, intertransverse fusion technique described by Moe (1958). There was then a deep and thorough decortication of all the posterior bony structures from the midline out to the tips of the transverse processes. This was followed by the application throughout the fusion area of large amounts of fresh autogenous iliac bone which had been cut into matchsticks. On average 11 vertebrae were fused (range 10 to 15 vertebrae). Figure 1 shows the apex of the curves, the side of the convexity and the vertebrae fused.

Post-operative care

After the operation all the patients were nursed free in bed for 7 to 10 days. An underarm plaster jacket was then

applied with the patient lying on the spinal table but without traction. The jacket was carefully moulded over the iliac crests and rib hump to support the curve. The patients were then immediately mobilised and allowed home within a few days. They returned to normal school and activities within a few weeks but were not allowed to participate in contact sports.

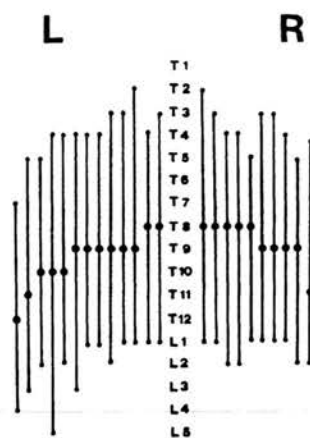


Fig. 1

Apex of curves, side of convexity and vertebrae fused.

According to a policy in earlier years, but now discontinued, 11 patients had the spine explored surgically after six months and all were found to have a solid fusion. After ten months the underarm jacket was removed and all patients were allowed unrestricted activities without external support.

Spinal assessment

The patients were routinely reviewed and radiographed when the plaster jacket was removed, then at one year, at 18 months and at 27 months after the operation. Thereafter they were seen at yearly intervals until the iliac apophyses were fully formed and fused and the spine was considered to be skeletally mature (Zaousis and James 1958).

All of the patients were specially recalled for the purpose of this investigation. Their spines were examined and the standing and sitting heights were recorded. Clinical photographs were taken and compared with those previously taken before operation and one year after operation. Antero-posterior and lateral radiographs of the erect spine were taken with the tube centred over the middle of the fused area.

Radiographic measurements of the fused region

The severity of the scoliosis was measured by the Cobb method on the erect antero-posterior spinal radiographs taken before operation, after the plaster jacket had been applied 7 to 10 days after operation, when the plaster jacket was removed 10 months after operation, and at

the final follow-up. Care was taken always to measure the Cobb angles from the same levels at either end of the fused region in each patient. Serial measurements of the fused region in each patient were made as follows. First, the length of the fusion in the longitudinal axis from the mid-point on the upper surface of the vertebra at the upper end of the fusion to the mid-point on the lower surface of the vertebra at the lower end of the fusion

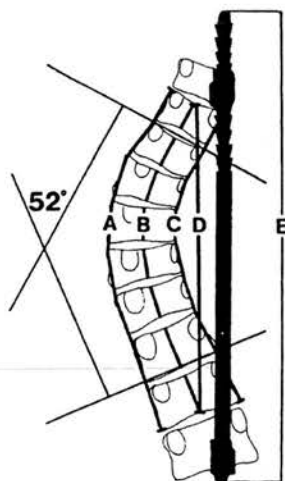


Fig. 2

Measurements on the antero-posterior radiographs. A: Distance along the convex side. B: Sum of the lengths of the vertebral bodies in their midportion. C: Distance along the concave side. D: Length in the longitudinal axis. E: Length of the Harrington rod (= reference distance).

(distance D in Fig. 2) was measured. Secondly, the distances along the convex (distance A in Fig. 2) and concave (distance C in Fig. 2) borders of the vertebrae in the fusion region were measured. In addition, the sum of the lengths of the vertebral bodies in their mid portion (distance B in Fig. 2) was calculated in the fused region. In order to overcome errors due to the differing magnifications of the serial radiographs, all the measurements were calibrated against the length of the Harrington rod (distance E in Fig. 2), which served as a reference distance for each patient. The radiographic difference in the length of the Harrington rod in each of the serial radiographs with reference to the first post-operative radiograph was used to calculate the degree of magnification, and the corresponding corrections were made to the distances measured on each radiograph. Errors due to magnification could be as large as 9 per cent, especially when comparing radiographs in plaster and without. We also compared the length of the fused region in the longitudinal axis (distance D in Fig. 2) on the post-operative radiograph with the same measurement on the

pre-operative radiograph in order to estimate the increase in height produced by correcting the scoliosis. This measurement could not be corrected for magnification because of the absence of the Harrington rod on the pre-operative radiograph.

Changes in rotation were measured with reference to the apical vertebral body on the serial radiographs for each patient. The distance between the border of the

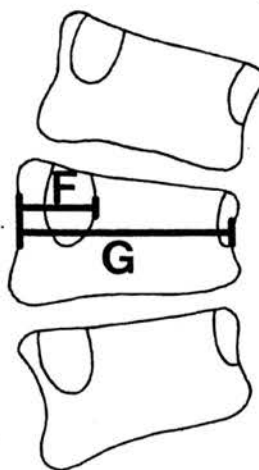


Fig. 3

Measurements of rotation. F: Distance between the border of the body on the convex side and the medial border of the pedicle on the same side. G: Width of the vertebral body at its narrowest site. The ratio F:G is calculated. On serial radiographs changes of this ratio on the same vertebral body result from changes in rotation.

body on the convex side and the medial border of the pedicle on the same side (distance F in Fig. 3) was measured. This was compared to the total width of the vertebral body at its narrowest site (distance G in Fig. 3) and the ratio F:G calculated. This ratio overcame errors in magnification and could be used for direct comparison on the serial radiographs.

RESULTS

There were no post-operative complications and all the patients developed a solid fusion. In 11 of the earlier patients the solid fusion was seen at routine surgical exploration performed six months after the fusion operation. This procedure was later discontinued because no pseudarthroses were found (McMaster 1980). Solid fusion was demonstrated on the antero-posterior, lateral and oblique radiographs of all of the patients taken when the plaster jacket was removed at 10 months.

Severity of scoliosis

Figure 4 shows the mean severity of the scoliotic curves and their range of values before operation, after appli-

cation of the plaster jacket seven to ten days later, at 10 months when the plaster jacket was removed, and at final follow-up. It should be noted that we measured the post-operative degree of severity on the erect spinal radiograph taken after the plaster jacket was applied. Occasionally there was a very slight loss of correction between the time of insertion of the Harrington rod and the application of the plaster jacket. We believe that the radiograph taken after the plaster jacket was applied was a more realistic measurement of the degree of correction

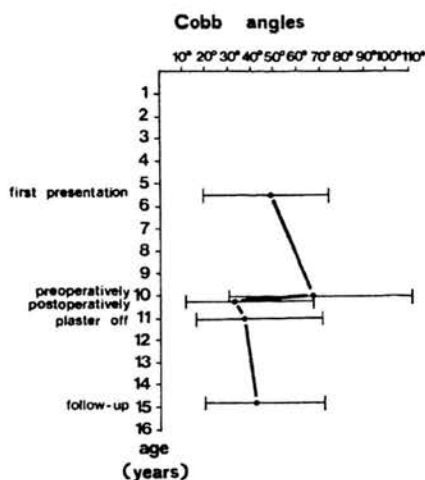


Fig. 4

Mean severity of the scoliotic curves and their range of values at first presentation, pre-operatively, seven days after operation (when plaster jacket was applied), 10 months after operation (when plaster was removed) and at final follow-up.

and a more suitable reference radiograph for the calculation of future loss of correction. The mean pre-operative curve was 66.9 degrees (range 30 to 112 degrees) and after operation this was reduced to a mean of 32.9 degrees (range 11 to 68 degrees). The resulting correction was 50.8 per cent or 34.0 degrees (range 11 to 54 degrees). Ten months after operation and out of plaster, the mean curvature was 37.0 degrees (range 16 to 72 degrees), and there was a mean loss of correction of 4.1 degrees or 6.1 per cent. At final follow-up, a mean 4.5 years after operation, the mean scoliosis was 41.7 degrees (range 20 to 72 degrees). The total loss of correction was now 8.8 degrees (range 2 to 19 degrees) or 13.2 per cent.

Severity of kyphosis

Fifteen patients had pre-operative lateral spinal radiographs and the mean degree of kyphosis in the area of the planned fusion was 28.7 degrees (range 10 to 60 degrees). This angle was not the same as the thoracic kyphosis: it was usually smaller because we measured only the vertebrae within the fusion area and not the whole of the thoracic spine. Five patients with infantile idiopathic scoliosis had a true kyphoscoliosis with an increased

kyphosis of over 40 degrees (range 44 to 64 degrees), within the planned fusion area. No patient had a pre-operative true thoracic lordosis. Ten months after operation and out of plaster, the mean degree of kyphosis for all the patients was 18.4 degrees (range 10 to 33 degrees) in the fusion area. There was, therefore, a mean reduction in the pre-operative kyphosis of 10.3 degrees (range 2 to 24 degrees) or 35.9 per cent. At final follow-up, a mean of 4.5 years after operation, the mean kyphosis was 18.1 degrees for all the patients. In five patients there had been no change, 12 patients had a reduction in the kyphosis of up to 10 degrees but were not lordotic, and six patients had an increase in kyphosis of between 5 and 24 degrees.

Longitudinal growth of the fusion area

The mean length of the fusion area in the longitudinal axis of the post-operative radiograph (distance D in Fig. 2) was 17.4 cm (range 12.9 to 21.6 cm). Ten months after operation there was a mean decrease in this distance, corrected for magnification, of 0.2 cm (range 0 to 0.6 cm) and by final follow-up this distance had increased only very slightly to a mean of 0.6 cm (range 0.1 to 1.5 cm). The mean longitudinal length of the fusion area was, therefore, 0.4 cm longer than that noted immediately after operation.

It was not possible to measure very accurately the increase in the longitudinal length of the fusion area before and after operation because the absence of a Harrington rod on the pre-operative radiograph prevented an assessment of the degree of magnification. An estimation of the difference in magnification on the post-operative radiograph was approximately 10 per cent, and with this correction the true increase in length produced by the operation was approximately 3 cm.

Vertebral body growth

In the first 10 months after operation the length (corrected for magnification) along the margins of the vertebral bodies and intervertebral discs on the convexity of the curve in the fused area (distance A in Fig. 2) increased by a mean 0.3 cm (range zero to 1.0 cm). At final follow-up there was an additional mean increase of 1.6 cm (range 0.1 to 3.2 cm) giving a total mean increase of 1.9 cm (range 0.5 to 3.7 cm). The same measurements were made along the concave side of the vertebral bodies (distance C in Fig. 2), and during the first 10 months the mean increase was 0.2 cm (range zero to 0.6 cm). By final follow-up there was a mean additional increase of 0.3 cm (range zero to 0.8 cm), giving a total increase of 0.5 cm (range zero to 1.2 cm). This increase is almost identical to the increase in the longitudinal axis (distance D in Fig. 2).

Post-operatively the sum of the heights of the vertebral bodies (distance B in Fig. 2) compared to the total length of the spine including the intervertebral discs

in the fusion area (distance B plus disc spaces) was a mean of 82.3 per cent. After ten months this increased to a mean of 86.7 per cent, and at final follow-up it was 95.6 per cent. During the first ten months the mean sum of the heights of the vertebral bodies in the fusion area increased by 1 cm (range 0.4 to 1.8 cm), and during the follow-up period there was a further increase of 2.2 cm (range 0.5 to 5.7 cm), making a total of 3.2 cm. The mean yearly

operation with Harrington instrumentation initially caused a partial derotation of the scoliosis, but during the following years the rotation again increased to its original severity and in some patients became more severe. The degree of initial derotation and subsequent rerotation varied considerably between the patients, but the tendency was always the same. In four patients the quality of the radiographs did not allow accurate measurements and they were excluded.

Height of Edinburgh Children

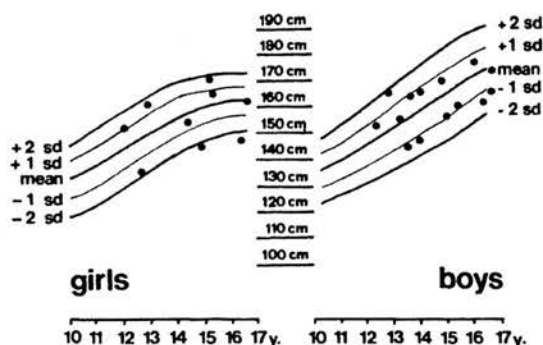


Fig. 5

Height of the 23 patients (9 girls and 14 boys) compared with the average height of Edinburgh children (standards of Department of Child Life and Health of the University of Edinburgh). The second standard deviation (± 2 sd) corresponds approximately to the third percentile.

increase in the height of a vertebral body in the fusion area was, therefore, 0.067 cm and this was accommodated by narrowing of the intervertebral disc spaces. In two patients the quality of the radiographs did not allow accurate measurements and they were excluded.

The mean width of the apical vertebral body after operation was 32.8 mm (range 27 to 40 mm) measured at its narrowest site, and 36.3 mm (32 to 43 mm) corrected for magnification on the radiograph at final follow-up. The mean increase in width of the vertebral body was, therefore, 3.5 mm.

Vertebral rotation

The rotation accompanying the scoliosis was assessed by a modification of the method described by Nash and Moe (1969). In our method the rotation of the apical vertebral body was expressed as a percentage by comparing the distance between the border of the pedicle on the convex side and the border of the body on the same side to the total width of the vertebral body (Fig. 3). This percentage does not represent the actual number of degrees of rotation but can be used to show relative changes in rotation for a given patient when the measurements are made on the same vertebral body on different occasions. The mean rotation was 38.6 per cent (range 20 to 55 per cent) before operation, 33.5 per cent (range 18 to 43 per cent) after operation and 39.3 per cent (range 28 to 47 per cent) at final follow-up. These findings show that the correction of the scoliosis at the time of

Standing and sitting heights

The standing heights of all but two of the patients at final follow-up were within the normal range for British children of an equivalent age (Tanner *et al.* 1966) (Fig. 5). Only two girls had standing heights below the third percentile and both these patients had small parents. The mean percentage of sitting to standing heights for all the patients was 50.8 per cent (range 42.5 to 53.8 per cent).

Illustrative case

A typical illustration case is shown in Figures 6-15.

DISCUSSION

A previous study from this Unit of children with idiopathic scoliosis fused posteriorly at 10 years of age showed that even a solidly fused spine continued to bend during the adolescent growth spurt, losing an average of 17 degrees of correction before becoming stable (McMaster and Macnicol 1979). Winter (1971) strongly advised supporting such young fusions with a Milwaukee brace to prevent bending during this period of rapid growth. Following these earlier findings it was decided by one of us to try to produce a more stable fusion by applying an additional massive onlay of autogenous iliac bone in an attempt to produce a wide and thick mass of bone which would be of sufficient strength to resist bending during the adolescent growth spurt. A single Harrington distraction rod used in combination with this fusion technique corrected the scoliosis by a mean of 51 per cent (mean 34 degrees), and all the spines developed a solid fusion. An underarm plaster jacket was worn for 10 months after operation while the fusion was becoming solid, and during this time there was a mean loss of correction of 4 degrees.

When the plaster jacket was removed the spine was left without support, and after a mean follow-up of 4.5 years there was an additional mean loss of 5 degrees despite the presence of the solid fusion. It is debatable whether this loss of correction is sufficiently severe to warrant the inconvenience of a Milwaukee brace, which would have have to be worn for several years after removal of the plaster jacket and might not be effective in stabilising the fusion.

The few previous studies of the growth of the spine after early posterior fusion have been based on the measurements of serial spinal radiographs (Ponseti and

Friedman 1950; Risser *et al.* 1966; Winter 1971; Letts and Bobechko 1974). These studies suffer from the inaccuracy caused by varying radiographic magnifica-

overcome by calibrating our measurements against the length of the Harrington rod, which is a known constant on the serial radiographs for each patient. Our corrected

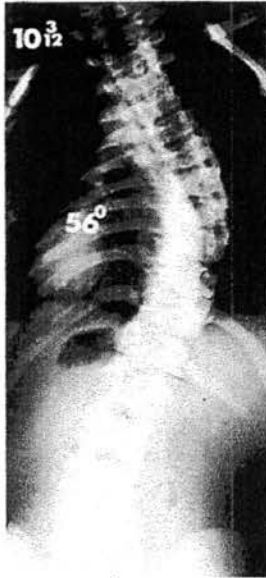


Fig. 6

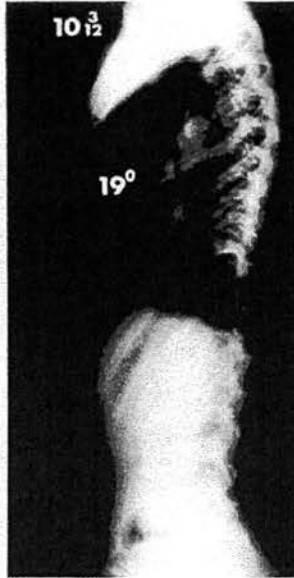


Fig. 7



Fig. 8

Case 1. Figure 6—Boy aged 10 years 3 months with severe juvenile idiopathic right thoracic scoliosis. The pre-operative erect spinal radiograph shows the curve to measure 56 degrees. Figure 7—Before operation there is a thoracic kyphosis of 19 degrees in the region of the proposed spinal fusion. Figure 8—There is a marked rib hump on the right side.

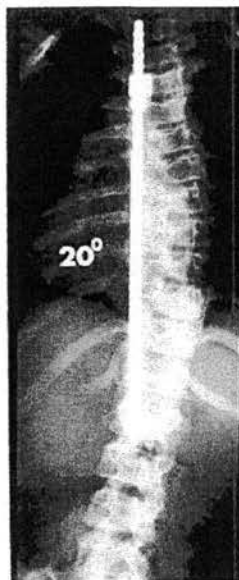


Fig. 9



Fig. 10



Fig. 11

Case 1. Figure 9—Ten months after posterior spinal fusion the radiograph shows good correction and solid fusion. The curve measures 20 degrees. Figure 10—After operation the kyphosis has been reduced to 11 degrees. Figure 11—Good cosmetic results; rib hump reduced.

tions of the spine produced by the differences in the distance of the patient from the radiographic plate on each occasion. In our study this inaccuracy has been

serial measurements, taken over a mean period of four and a half years after solid fusion, showed no longitudinal growth in the posteriorly fused elements. This confirms

the findings of Winter (1971) and of Letts and Bobechko (1974) but contradicts the observations of Risser *et al.* (1966).

Although the posterior fusion mass did not grow longitudinally, the vertebral bodies in the fused region continued to increase in height and width. The normal growth in height of one vertebral body is estimated to be 0.07 cm per year (Moe *et al.* 1978). In our patients, the growth in height of the vertebral bodies in the fused region was 0.067 cm per year and was therefore not significantly impaired by the fusion. This increase in height was presumably due to a continuation of the

straighten the kyphosis as the lateral curvature was corrected (Fig. 10). After operation the thoracic kyphosis was reduced by a mean of 10.3 degrees, but on average there was no further reduction during the period of follow-up. Whereas most patients showed a mild further reduction of the kyphosis or remained unchanged, a few had an increase of kyphosis in spite of the growth of the anterior elements. We believe that the thick posterior fusion was of sufficient strength to prevent a significant reduction in the kyphosis or the development of a lordosis despite the tension produced by the continued longitudinal growth of the vertebral bodies anteriorly.

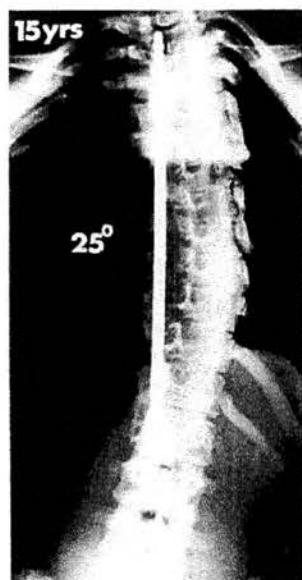


Fig. 12



Fig. 13



Fig. 14



Fig. 15

Case 1. Figure 12—Radiograph taken at the age of 15 years shows that there has been a slight loss of correction and the curve now measures 25 degrees. The intervertebral disc spaces in the region of the spinal fusion are markedly narrowed and there has been a recurrence of the vertebral rotation. Figure 13—The intervertebral disc spaces in the region of the fusion have narrowed considerably and there has been a slight loss in the post-operative degree of kyphosis. Figure 14—Cosmetically there is still a good result. Figure 15—The forward bending view shows that the rib hump has returned.

normal process of epiphyseal growth at the end plates on the upper and lower surfaces of the vertebral bodies (Bick and Copel 1950). In our study the increase in height of the vertebral bodies in the posteriorly fused region was initially accommodated by narrowing of the intervertebral disc spaces—a finding previously noted by Risser *et al.* (1966). However, despite the narrowing of the intervertebral disc spaces there was no case of spontaneous interbody fusion. In animal experiments continued anterior growth of the vertebral bodies produced an increasing lordosis following posterior fusion (Veliskakis and Levine 1966; Coleman 1968). Bonnett *et al.* (1975) also noted a lordosis developing after long posterior fusions in young children with poliomyelitis. Before operation all of our patients had some degree of thoracic kyphosis, and four patients with infantile idiopathic scoliosis had a greater than normal kyphosis in the upper thoracic region. When the Harrington distraction rod was applied during operation it tended partially to

Our measurements have shown that during the follow-up period there was an increase of up to 2 cm in length of the convexity of the curve along the lateral margin of the vertebral bodies, with no corresponding increase on the concave side. It was also found that immediately after Harrington correction there was a partial reduction in the degree of vertebral rotation but that during the follow-up period the rotation again returned to the same severity or more. No measurements were made of the rib hump, but comparison of the clinical photographs taken before operation, when the plaster jacket was removed and at final follow-up showed that although the rib hump was reduced after operation (Fig. 11) it recurred as the patient grew (Fig. 15). In our opinion, the increase in length on the convexity of the curve and the recurrence of vertebral rotation are due to the continued longitudinal growth of the vertebral bodies. Once the degree of accommodation provided by the intervertebral disc spaces has been taken up, the vertebral

bodies bulge laterally towards the convexity of the curve and pivot on the unyielding posterior fusion mass. It is likely that this contributes to the post-operative loss of correction and causes recurrence of the rib hump despite the presence of a solid posterior fusion. The increasing rib hump gives the illusion of a developing thoracic lordosis, but this is not confirmed on the lateral radiographs. So far as we know these observations have not been previously reported. These changes are not so apparent in patients with adolescent scoliosis, who are usually girls and undergo fusion after the age of thirteen years when the adolescent growth spurt is well advanced.

A criticism of early posterior spinal fusion is that because it prevents further longitudinal growth in the fused region it will produce a relative shortening of the spine as the child grows to maturity. Winter (1977) has produced a formula from standard growth tables which gives the approximate potential shortening of the spine caused by the fusion: $\text{centimetres of shortening} = 0.07 \times \text{number of segments fused} \times \text{number of years of growth remaining}$. In our patients the mean number of segments fused was 11, and assuming six years of growth remaining, the anticipated shortening caused by the spinal fusion would have been 4.6 cm. It should, however, be remembered that the straightening of the spine at operation increased the overall height by approximately 3 cm. Also if the spine had not been corrected and fused,

the amount of shortening caused by the progressive curve is likely to have been greater than the shortening caused by the fusion. Although only six of our patients had reached skeletal maturity, the great majority had progressed sufficiently through the adolescent growth spurt to the stage where the growth rate was decelerating. At final follow-up only two of the 22 children were below the third percentile for the average height for British children of an equivalent age. However, on inspection the majority of children did appear to have mild to moderate stunting of the spine, and the mean percentage of sitting to standing height was 50.8 per cent, which is below the normal range of 52 to 53 per cent at skeletal maturity (Moe *et al.* 1978). This stunting of the spine is considered acceptable in comparison with the degree of deformity which would have occurred had the spine not been fused.

In conclusion we can state that although a posterior spinal fusion stops the longitudinal growth in the posterior elements, the vertebral bodies continue to grow anteriorly and this is most apparent during the adolescent growth spurt. A thick posterior fusion mass prevents the development of a lordosis but the anterior growth causes the vertebral bodies to bulge laterally towards the convexity and also to pivot on the posterior fusion, giving rise to a loss of correction, increasing vertebral rotation and recurrence of the rib hump.

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Section E

Spinal Surgery

PSEUDARTHROSIS AFTER SPINAL FUSION FOR SCOLIOSIS

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A series of 246 patients with scoliosis and attempted fusion had exploration performed six months later in order to detect and treat any pseudarthrosis at an early stage and so prevent subsequent loss of correction. Bilateral or unilateral pseudarthroses occurred in 25 per cent and were of three types—definite, hairline and doubtful. Single unilateral pseudarthroses accounted for 6 per cent and were of little if any clinical significance. The hairline pseudarthroses could not be seen radiologically and were easily missed at exploration. In general the pseudarthroses were least common in the more rigid parts of the spine and in curves which by nature of their aetiology or long duration had become most rigid. Neither the initial severity of the curve nor the degree of correction obtained before the initial attempted fusion had any apparent effect on the incidence. Follow-up for an average of four years has shown that a pseudarthrosis is of little significance with regard to the ultimate result provided it is recognised early and repaired.

The major cause of deterioration in a scoliotic curve following attempted spinal fusion is a pseudarthrosis; the curve may relapse to its former severity or even worse if the patient is still growing (Risser 1964).

The reported incidence of pseudarthrosis using radiographs for detection has varied considerably; without internal fixation it has varied from 3.3 to 68.3 per cent (Ponseti and Friedman 1950; Cobb 1952; Gucker 1956; Blount, Schmidt, Keever and Leonard 1958; Risser and Norquist 1958; Moe 1958; Goldstein 1959; Moe and Gustilo 1964; Winter, Moe and Eilers 1968; Pavon and Manning 1970), with an average of 22.5 per cent. Following the introduction of Harrington instrumentation the reported incidence based on radiographic findings alone has declined and now ranges from 2 to 17 per cent (Winter *et al.* 1968; Moe and Valuska 1968; Goldstein 1969; Hall and Gillespie 1971; Hall and Spira 1973; Leider, Moe and Winter 1973; Dickson and Harrington 1973), with an average of 6.4 per cent.

Cobb (1952) has stated that the only way to be accurate in a study of pseudarthrosis is to explore each spine, and even then one may be overlooked. Few surgeons have done this routinely at an early stage (Outland, McDowell and Flynn 1964; May and Mauch 1966; Ashley 1967; Graham 1968; Donaldson, Wissinger and Stone 1969; Mathews and Stelling 1970). These series are relatively small but have revealed a much higher incidence, namely, 11 to 69 per cent with an average of 42.3 per cent.

James (1965) reported that his earlier review of 400 patients with scoliosis treated before 1958 was of little value because radiographs were unreliable. Since 1958 it has been his policy to explore all fusion masses at six months. The purpose of this paper is to report the findings and to discuss the various factors that may influence the development of a pseudarthrosis.

CLINICAL MATERIAL

At the Princess Margaret Rose Orthopaedic Hospital, Edinburgh, during the period 1962 to 1973, 246 patients with scoliosis had exploration performed six months after the attempted fusion. There were 162 girls and eighty-four boys, whose ages at spinal fusion ranged from three to twenty-eight years (Fig. 1). One hundred and twenty patients had idiopathic scoliosis, sixty-five had congenital scoliosis and forty-six paralytic scoliosis (Table I). Of the fourteen patients in a miscellaneous group seven had neurofibromatosis, two cerebral palsy, two Kugelberg-Welander syndrome, two Marfan's syndrome and one amyotonia congenita.

Correction before operation in this series was by Risser localiser jacket in 242 patients. In twenty-five patients further correction was obtained by means of

TABLE I
FINDINGS AT EXPLORATION OF 246 SPINAL FUSIONS

Aetiology of scoliosis	Number of patients	Solid fusion	Number of pseudarthroses and incidence (per cent)	
			Bilateral	Single unilateral
Idiopathic				
—adolescent	64	44	17 (26.5)	3 (5)
—juvenile	35	25	4 (11)	6 (17)
—infantile	22	19	3 (14)	0 (0)
Congenital	65	54	7 (11)	4 (6)
Paralytic	46	32	13 (28)	1 (2)
Miscellaneous	14	11	3 (21)	0 (0)
Totals	246	185	47 (19)	14 (6)

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Harrington instrumentation, a technique now used routinely for the more severe curves. Halo-pelvic traction (O'Brien, Yau and Hodgson 1973) was used in four patients.

Spinal fusion was performed by the technique described by Moe (1958), in which small carefully-shaped

If a pseudarthrosis was discovered it was excised leaving a trench in the fusion mass about 7 millimetres wide over the length of the defect but not down to ligamentum flavum (Fig. 2). The walls were undercut to expose bleeding cancellous bone. A small carefully-shaped block of cortico-cancellous bone from the iliac

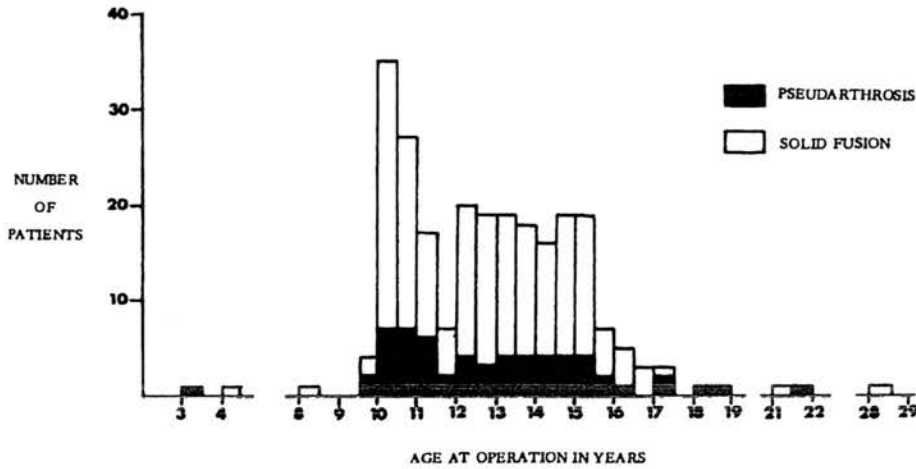


FIG. 1

A histogram relating the age at operation to the number of patients developing a pseudarthrosis.

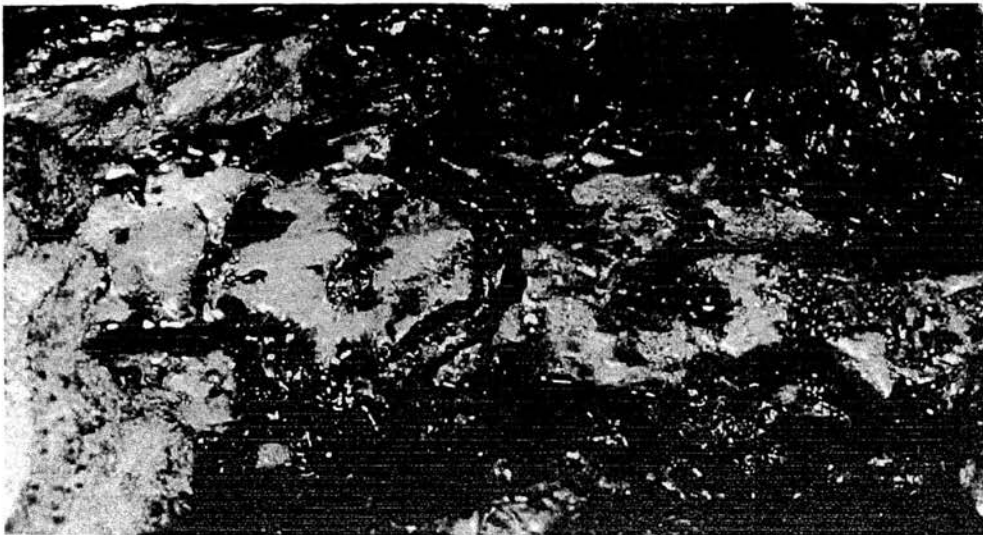


FIG. 2

A photograph taken after a bilateral definite pseudarthrosis had been excised leaving a trench across the fusion mass.

blocks of iliac cortico-cancellous bone are punched into the excised interfacetal joints. After operation the patient was kept in bed for four weeks before being allowed home in his Risser jacket. At six months the spine was explored; if solidly fused, the patient wore a Milwaukee brace continuously for six months.

crest was gently impacted into the trench, leaving the cortical surface of the graft just beneath that of the fusion mass (Fig. 3). The patient was then placed in a new Risser localiser jacket. After five months re-exploration limited to the area of repair was performed, and if solid a Milwaukee brace was worn for six months.

FINDINGS AT EXPLORATION OF THE FUSION MASS

Usually the periosteum strips easily, exposing a sheet of cortical bone which extends between the neutral vertebra above and below and out to the tips of the transverse processes in the thoracic region (Fig. 4). Often the only recognisable structures are the spinous processes, now just small rounded projections between which may be

partially across the fusion mass. Any movement at the pseudarthrosis is minimal, and unless all the soft tissue is carefully cleared it will be missed. Histological examination has shown that the crack contains fibrocartilage.

Thirdly there is the *doubtful* pseudarthrosis. By this we mean an area in which the periosteum is more adherent than in adjacent areas and the cortical bone appears soft. If this bone is curetted away a defective area in the cancellous bone is exposed.



FIG. 3

Same case as in Figure 2. Blocks of cortico-cancellous bone from the iliac crest have been punched into the trench.

small gaps representing the interlaminar spaces. In the lumbar region the bony anatomy is not quite so completely obliterated and the sites of the interfacetal joints can often be located.

The first suspicion of a pseudarthrosis comes during the stripping of the periosteum. If it does not strip easily and is bound to the underlying bone by fibrous tissue, that area should be carefully inspected for the presence of a pseudarthrosis. Starting from the top of the fusion mass, each segment of the vertebral column is meticulously cleared of all soft tissue. The spinous processes of neighbouring vertebrae are grasped by sequester forceps and any movement, however small, is noted.

Types of pseudarthrosis—There are three basic types. Firstly, there is the *definite* pseudarthrosis, which is the most common type and presents as an irregular crevice filled by fibrous tissue (Fig. 5). The crevice may extend completely across the fusion mass (bilateral pseudarthrosis), when motion between the vertebrae is easily elicited, or only part of the way across either on the concave or convex side of the curve (unilateral pseudarthrosis), with a solid bar of bone on the other side.

Secondly, there is the *hairline* pseudarthrosis, which is much less common and very difficult to see at exploration. Again it may extend either completely or only

Patients with pseudarthrosis—Fifty-four out of the 246 patients were found to have a pseudarthrosis at exploration. Seven patients thought to have a solid fusion at revision later developed increasing deformity and required re-exploration. This was performed from twelve to thirty-three months (average twenty-one months) after the original attempt at fusion, and all seven patients were found to have a pseudarthrosis. Three had only a single bilateral hairline pseudarthrosis, which despite a rather innocuous appearance had already given rise to a serious loss in correction (average 21 degrees). Of the remaining four patients, three had a single bilateral definite pseudarthrosis and one had two bilateral definite pseudarthroses.

The total number of patients developing a pseudarthrosis was therefore sixty-one, giving an overall rate of 25 per cent. Fourteen of these patients had only a single unilateral pseudarthrosis of doubtful importance. Therefore if we consider only the forty-seven patients with bilateral pseudarthroses, the rate is 19 per cent (Table I).

Number of pseudarthroses—Thirty-seven of the sixty-one patients had a pseudarthrosis at only one level, fourteen at two, five at three, four at four and one at seven levels. This gives a total of 103 pseudarthroses after 2,040

attempts at fusion at the various vertebral levels. The overall risk of pseudarthrosis at any one level was therefore 5 per cent.

Site of the pseudarthroses—The distribution of the various types of pseudarthrosis is shown in Figure 6. There were fifty-eight definite bilateral pseudarthroses. There were

sixty-one patients who had their pseudarthroses repaired, thirty-five underwent re-exploration three to fourteen months later (average six months). This is now done routinely.

Six patients were found to have a single bilateral definite pseudarthrosis which had remained unhealed, but

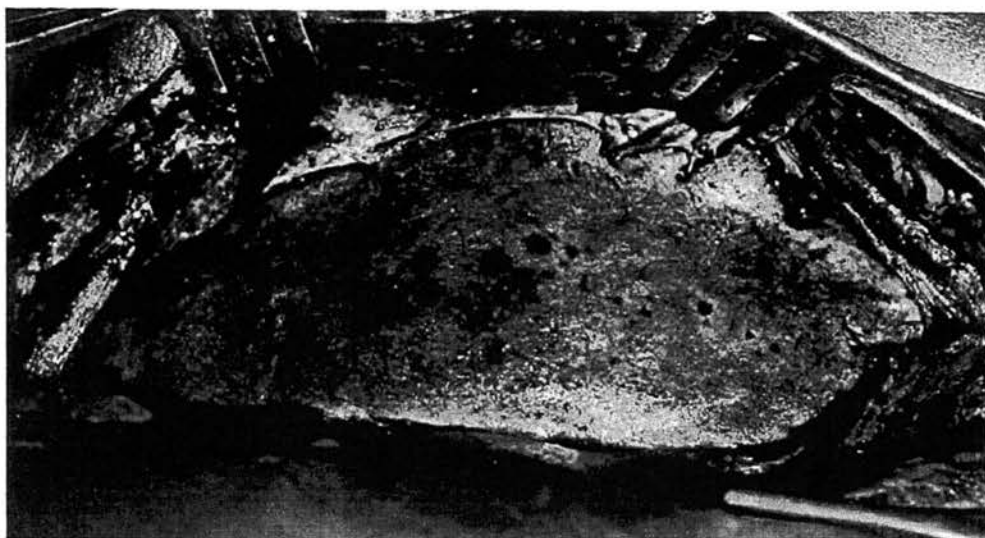


FIG. 4

A photograph taken at exploration showing solid fusion in the thoracic region.



FIG. 5

In this photograph two definite bilateral pseudarthroses in the thoracic region are clearly seen.

thirty-seven definite unilateral pseudarthroses, twenty-two on the convex side and fifteen on the concave side. There were four hairline pseudarthroses, all bilateral. Four pseudarthroses were of the doubtful variety, two on the concave side and two bilateral.

Re-exploration of the repaired pseudarthroses—Of the

the remaining twenty-nine had solid fusion. Of the six unhealed pseudarthroses, two were between T.9 and T.10, two between T.12 and L.1 and two in the lumbar region. All six were again repaired as before. Only one spine was re-explored for a second time five months after the second repair and had soundly fused.

FOLLOW-UP

The 246 patients were followed for periods varying from one to sixteen years (average four years). The average follow-up for the thoracic, thoraco-lumbar and double curves of each aetiology is shown in Tables II, III and IV, along with the loss in correction during and after the first year. No patient was dismissed until skeletally mature.

Loss of correction in the first year—In those patients whose spinal fusion was found to be solid the average loss of correction in the first year for a thoracic curve was 5 degrees and for a thoraco-lumbar curve 6 degrees. The greatest loss occurred in paralytic scoliosis and the least in congenital scoliosis.

Aetiology of the scoliosis (Table I)—The lowest incidences occurred in infantile idiopathic scoliosis (14 per cent) and congenital scoliosis (17 per cent). These curves by nature of their long duration are often the most rigid types of scoliosis and therefore we think the most likely to proceed to solid fusion.

Two of seven patients with neurofibromatosis and one of two patients with Marfan's syndrome developed a pseudarthrosis but these numbers are too small for statistical analysis. Of the six patients found to have a pseudarthrosis still present after attempted repair, three had adolescent idiopathic scoliosis, two congenital scoliosis and one Marfan's syndrome.

Site of the curve—The only level at which a pseudarthrosis did not occur was between T.2 and T.3. Fusion

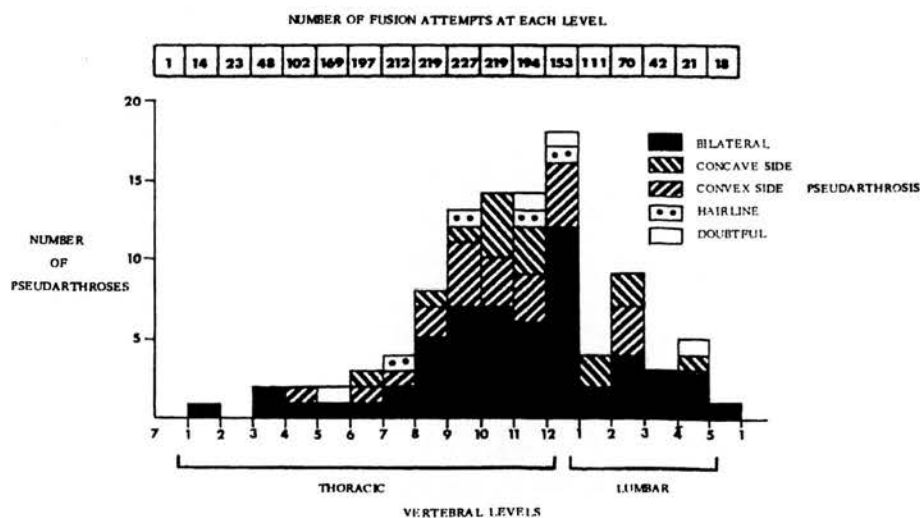


FIG. 6

A histogram showing the number and the type of the pseudarthroses at each vertebral level.

In those patients who were found to have a pseudarthrosis the average loss for a thoracic curve was 7 degrees and for a thoraco-lumbar curve 9 degrees.

Loss of correction after the first year—Both those patients with a solid fusion and those who had had a pseudarthrosis repaired lost a little correction after the Milwaukee brace was removed. In those with sound fusion the thoracic curves lost on average 7 degrees and the thoraco-lumbar curves 4 degrees.

In those patients with a proven pseudarthrosis which had been repaired the thoracic curves lost on average 6 degrees and the thoraco-lumbar curves 8 degrees. The greatest loss occurred in paralytic scoliosis.

FACTORS INFLUENCING THE DEVELOPMENT OF A PSEUDARTHROSIS

Age and sex—No one age group was found to be more susceptible (Fig. 1) and the distribution between the sexes was equal.

at this level, however, was attempted only twenty-three times. The majority were found between T.6 and L.3, where the greatest number of fusion attempts took place (Fig. 6). However, if we compare the number of pseudarthroses at each vertebral level and compare this with the number of times fusion was attempted at that level we see that the highest incidence of pseudarthroses occurred from T.9 to S.1 and the lowest in the mid-thoracic region (Fig. 7).

The vertebral levels with the highest rates of pseudarthrosis were in the more mobile part of the spine. This mobility we think is probably a predisposing factor.

Unilateral pseudarthroses were more common on the convex side of the curve where the grafted areas were subject to a distracting force and therefore we think more at risk.

Initial severity of the curve—The size of the initial curve before correction had no apparent effect (Tables II, III and IV).

Degree of correction—There was no significant difference

in the degree of correction obtained in those spines which developed a pseudarthrosis and those that did not (Tables II, III and IV). Risser and Norquist (1958), however, found pseudarthrosis more common in patients with the most correction.

Technique of fusion—The interfascial type of fusion popularised by Moe (1958) gives fewer pseudarthroses

spine. In our series seven out of twenty-five patients with Harrington instrumentation were affected (28 per cent). Graham (1968) explored forty-five patients with idiopathic scoliosis one to two years after a spinal fusion with Harrington rod correction and found an incidence of 55.5 per cent. In our experience the presence of a Harrington rod does not seem to lower the pseudarthrosis

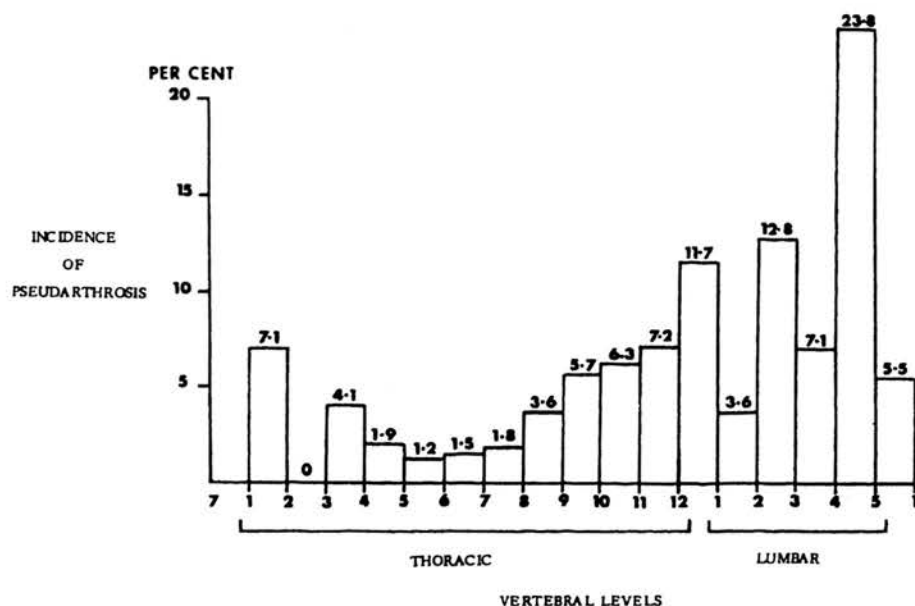


FIG. 7

A histogram showing the incidence of pseudarthrosis at each vertebral level.

TABLE II
DETAILS OF 120 CASES OF IDIOPATHIC SCOLIOSIS (INFANTILE, JUVENILE, ADOLESCENT)

Solid fusion							Pseudarthrosis						
Type of curve	Number of patients	Average curve pre-op. (degrees)	Average curve post-op. (degrees)	Average loss in correction (degrees)		Average follow-up (years)	Type of curve	Number of patients	Average curve pre-op. (degrees)	Average curve post-op. (degrees)	Average loss in correction (degrees)		Average follow-up (years)
				At 1 year	After 1 year						At 1 year	After 1 year	
Thoracic	68	65 (24-124)	38	6	8	4	Thoracic	17	65 (36-104)	39	9	6	3½
Thoracolumbar	16	65 (27-105)	35	8	5	4	Thoracolumbar	17	55 (33-83)	27	11	5	3
Double	2	68 68	34 34	10 12	3 6	3	Double	0	—	—	—	—	—

(Blount *et al.* 1958). We have not found it necessary to decorticate the bone or to add bone grafts because we feel that the careful stripping of the periosteum and meticulous clearance of all soft tissues provide an osteogenic stimulus in the growing child.

Harrington instrumentation is said to have helped lower the incidence of pseudarthrosis by stabilising the

rate, although the support offered by the rod may prevent the loss of correction which is often the only indication of a pseudarthrosis, and once the spine stops growing the tendency to lose correction becomes very much less.

Plaster jacket immobilisation—The majority of our patients were allowed to walk at four weeks in a localiser jacket. The use of localiser jackets is said to have lowered

TABLE III
DETAILS OF SIXTY-THREE CASES OF CONGENITAL SCOLIOSIS

Solid fusion							Pseudarthrosis						
Type of curve	Number of patients	Average curve pre-op. (degrees)	Average curve post-op. (degrees)	Average loss in correction (degrees)		Average follow-up (years)	Type of curve	Number of patients	Average curve pre-op. (degrees)	Average curve post-op. (degrees)	Average loss in correction (degrees)		Average follow-up (years)
				At 1 year	After 1 year						At 1 year	After 1 year	
Thoracic	32	54 (33-120)	31	3	5	4	Thoracic	6	57 (41-78)	40	4	7	6
Thoracolumbar	19	67 (15-102)	42	3	4	4	Thoracolumbar	4	66 (51-89)	37	12	9	4
Double	1	73 100	60 82	0 0	— —	1	Double	1	64 47	40 18	4 26	16 0	6

TABLE IV
DETAILS OF FORTY-SIX CASES OF PARALYTIC SCOLIOSIS

Solid fusion							Pseudarthrosis						
Type of curve	Number of patients	Average curve pre-op. (degrees)	Average curve post-op. (degrees)	Average loss in correction (degrees)		Average follow-up (years)	Type of curve	Number of patients	Average curve pre-op. (degrees)	Average curve post-op. (degrees)	Average loss in correction (degrees)		Average follow-up (years)
				At 1 year	After 1 year						At 1 year	After 1 year	
Thoracic	17	70 (30-120)	40	6	6	4	Thoracic	2	76 (62-91)	45	5	5	2
Thoracolumbar	14	88 (25-115)	46	11	1	5	Thoracolumbar	12	77 (54-113)	47	5	13	3
Double	1	84 60	46 30	11 8	3 28	7½	Double	0	—	—	—	—	—

the incidence of pseudarthrosis (Risser and Norquist 1958; Brown 1965) and to have allowed earlier mobilisation of the patient without apparent increase in the pseudarthrosis rate (Risser and Norquist 1958; Leider *et al.* 1973).

Ponseti and Friedman (1950) found that the pseudarthrosis rate was not influenced by the use of a cast for longer than five months. We have found, however, that even though a fusion was seen to be solid at six months the bone mass was not sufficiently mature to prevent bending and required further protection for six months in a Milwaukee brace. Even when the brace was removed at one year a solid fusion mass would bend a little, causing a slight loss of correction before becoming stable.

DISCUSSION

This study has shown that a pseudarthrosis after spinal fusion for scoliosis is a not uncommon finding when every spine is explored at six months. Most surgeons do not

do this, but rely on various oblique radiographic projections to detect a pseudarthrosis. Unfortunately these radiographs are often equivocal. A hairline pseudarthrosis cannot be seen radiographically. This type is seen only at exploration and even then it is easily missed, as were three in our series.

A rapid loss of correction of over 15 to 20 degrees is generally accepted as an indication of a pseudarthrosis, which may or may not be visible radiographically. The seven patients in our series with a pseudarthrosis which had been missed at exploration presented in this manner. James (1965) found that it was easier to prevent the loss of correction due to a pseudarthrosis by routine re-exploration and repair if necessary, rather than to wait and try to regain correction once it had been lost.

The follow-up in the present series has shown that in the first year both those patients with a solid fusion and those who had had a pseudarthrosis repaired lost a little correction while still in a cast or Milwaukee brace. This loss was on average 3 degrees greater in patients

with a repaired pseudarthrosis. After the first year there was again a slight loss in correction after the Milwaukee brace had been removed and while the fusion was maturing, and this was on average 2 degrees greater when a pseudarthrosis had been repaired. From these findings we see that the final difference in the amount of correction

lost between curves which had been found solid and those with a pseudarthrosis successfully treated is small, being of the order of 5 degrees. Therefore we can say that the presence of a pseudarthrosis is of little significance with regard to the ultimate result provided it is recognised early and repaired.

ADDENDUM

Nineteen surgeons of all grades of experience were involved in fusing these 246 spines. During the year 1975-76 sixty-two spines were fused by the two authors. Forty-four of these patients had Harrington rod instrumentation. All of these spines have now been re-explored and a single pseudarthrosis was found in only two patients, giving a pseudarthrosis rate of 3.2 per cent. One was a hairline pseudarthrosis found between the twelfth thoracic and first lumbar vertebrae in a long idiopathic thoraco-lumbar curve, and the other a definite bilateral pseudarthrosis at the lumbo-sacral junction in a patient with neurofibromatosis. Both of these patients had Harrington rod instrumentation.

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STABILITY OF THE SCOLIOTIC SPINE AFTER FUSION

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The factors during and after operation which influence the development of a solid and stable posterior spinal fusion have been evaluated in 406 patients with scoliosis. The patients were managed in three different ways and all pseudarthroses were accurately detected by exploring the spines six months after the attempted fusion. The incidence of pseudarthroses was significantly lowered from 25 per cent in Group I to 3.8 per cent in Group III by the application of Harrington instrumentation and the use of large amounts of autogenous iliac bone grafts in addition to an interfacetal fusion. Early mobilisation 7 to 10 days after operation and a return to normal activities in a well-moulded underarm plaster jacket did not have a detrimental effect on the development of the fusion or the early maintenance of correction. Those spines with supplementary bone grafts stabilised more rapidly and had better maintenance of correction with only minimal loss after removal of all external support at 10 months.

The stability of the scoliotic spine after fusion depends on producing a solid fusion of sufficient strength to resist bending under the influence of gravity and the deforming factors which first caused the curvature. Correction may be lost due either to the development of a pseudarthrosis or to bending of a solid but weak fusion. Serial spinal radiographs, measured using Cobb's technique (1948), are an excellent means of showing deterioration of the curve but are not always capable of distinguishing between a pseudarthrosis and bending of a solid fusion. Cobb (1952) stated that the only way to be absolutely accurate in the study of spinal fusions was to explore each spine surgically. Only a relatively few surgeons have done this routinely but all have found a much higher incidence of pseudarthroses than were apparent radiographically (Outland, McDowell and Flynn 1964; May and Mauck 1966; Ashley 1967; Graham 1968; Donaldson, Wissinger and Stone 1969; Mathews and Stelling 1970; McMaster and James 1976).

The purpose of this paper is to investigate the important operative and postoperative factors influencing the development of a solid and stable spinal fusion in patients with scoliosis. Three groups of patients treated by posterior spinal fusion but managed in different ways are compared, with special attention being given to the effects of Harrington instrumentation, the use of supplementary autogenous bone grafts and early mobilisation in a simple underarm cast. In order accurately to detect all pseudarthroses the spines were surgically explored six months after the attempted fusion. It was then possible to study radiographically the factors influencing the stability of the solid fusion.

CLINICAL MATERIAL

At the Princess Margaret Rose Orthopaedic Hospital, Edinburgh, during the period 1962 to 1977, 406 consecutive patients with scoliosis of the idiopathic, congenital and paralytic varieties have had a posterior spinal fusion followed by an exploration six months later. This second operation was carried out, as advocated by James (1965), to detect and repair pseudarthroses at an early stage and so prevent subsequent loss in correction.

The 222 patients with scoliosis operated on before 1973 have been studied retrospectively by reviewing their case notes and spinal radiographs taken before operation and during the follow-up period. The majority of these patients had their scoliosis corrected by the application of a Risser localiser jacket without Harrington instrumentation and have been the subject of a preliminary investigation into the incidence of pseudarthroses (McMaster and James 1976). Nearly all of these patients have now been followed to skeletal maturity and are used as a control group with which to compare the results obtained in the prospective study.

Since 1973 I have carried out a prospective study on a further 184 patients with scoliosis who have had a posterior spinal fusion followed by an exploration six months later. The majority of these patients were operated on by myself and had their scoliosis corrected using Harrington instrumentation with a specific attempt being made to lower the incidence of pseudarthrosis by modifying the fusion technique and the postoperative management.

Of the total number of patients 284 were girls and 122 boys, whose ages at spinal fusion ranged from 7 to 28 years (mean 12 years 5 months). The aetiology of the scoliosis in 239 patients was idiopathic, 98 were congenital and 69 neuropathic. Three hundred and eighty-four patients had a single structural curve of which 303 were thoracic, 78 thoracolumbar and three lumbar. There were 22 patients with a double structural scoliosis of which five were double thoracic and 17 had combined thoracic and lumbar curves.

METHODS

The patients fell naturally into three study groups, each group representing a step in the evolution of either the method of correction, technique of posterior spinal fusion or postoperative management (Tables I and II).

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Group I (243 patients). The scoliosis was corrected using a Risser localiser jacket followed one week later by a posterior spinal fusion through a large window cut in the back of the cast. No internal fixation was used and the spine was fused using only the interfacetal, intertransverse fusion technique described by Moe (1958). In this method of fusion the interfacetal joints are excised by turning back leaves of bone to come into contact in the intertransverse spaces. Small carefully shaped blocks of corticocancellous bone are then punched into the excised joints. The posterior elements were not decorticated nor were supplementary onlay bone grafts used. After operation the patient was kept in bed for one to three months before being allowed home in the Risser jacket which was not changed until exploration. Activities until exploration were confined to the house and garden.

Table I. Method of correction and technique of fusion

	Method of correction	Technique of fusion
Group I	Risser localiser jacket	A Moe interfacetal, intertransverse fusion alone
Group II	Harrington instrumentation	A Moe interfacetal, intertransverse fusion alone
Group III	Harrington instrumentation	A Moe interfacetal intertransverse fusion plus decortication of all posterior spinal structures and an onlay of autogenous iliac bone grafts

Group II (57 patients). The scoliosis was corrected using the Harrington distraction system. In 30 patients preliminary correction was obtained by means of a Risser localiser jacket applied one week before operation. As in Group I the spines were fused using the Moe (1958) interfacetal, intertransverse fusion technique without decorticating the posterior elements or adding supplementary onlay bone grafts. Immediately after operation anterior and posterior plaster shells were applied and maintained for two weeks. Then a simple

Table II. Postoperative management

	Bed rest		Plaster immobilisation	
	Type	Duration	Type	Duration (months)
Group I	Localiser jacket	1 to 3 months	Localiser jacket Milwaukee brace	6 } 12
Group II	Plaster shells	2 to 4 weeks	Underarm plaster jacket	12
Group III	Free in bed	7 to 10 days	Underarm plaster jacket	10*

*the first 31 patients in this group wore the plaster jacket for 12 months

underarm plaster jacket was applied with the patient lying on the Risser plaster table but without traction. The plaster was carefully moulded over the iliac crests and rib hump to support the curve. The first 20 patients were kept in bed for at least four weeks but the last 33 patients were allowed up after two weeks. Activities until exploration were again confined to the house and garden.

Group III (106 patients). These patients were studied prospectively and all the spinal fusions and subsequent explorations performed by myself. As in Group II the scoliosis was corrected using the Harrington distraction system but without preliminary traction or the application

of a Risser localiser jacket. The Moe (1958) interfacetal, intertransverse fusion technique was again used but in this group the posterior spinal elements were also thoroughly and deeply decorticated out to the tips of the transverse processes and a large number of fresh autogenous iliac bone grafts which had been cut into matchsticks were onlaid. After operation all patients were nursed free in bed for 7 to 10 days and then a simple well-moulded underarm plaster jacket was applied, as in Group II. They were then allowed home and encouraged to return to normal school and activities within a few weeks but were not allowed to participate in active sports.

At six months the patients in all three groups were readmitted to hospital where their plaster casts were removed and their spines explored. If solidly fused, the patients in Group I were placed in a Milwaukee brace for a further six months whereas patients in Groups II and III were replaced in their underarm plaster jackets. All of the patients in Group II and the first 31 patients in Group III wore an underarm jacket for an additional six months. Seventy-five of the more recent patients in Group III wore their jacket for only an additional four months.

RESULTS

Degree of correction. The number of degrees of correction and the percentage correction for the thoracic, thoracolumbar and lumbar curves in the three groups are shown in Tables III, IV and V. The greatest correction was obtained in Group III where the mean correction in the thoracic curves was 34 degrees (51 per cent), in the thoracolumbar curves 55 degrees (64 per cent), and in the lumbar curves 41 degrees (55 per cent).

Patients with pseudarthroses. The findings at exploration of the fusion in the three groups are shown in Table VI. Seventy-four of the 406 patients were found to have pseudarthroses at one or more levels. The lowest incidence of pseudarthroses occurred in Group III (3.8 per cent) followed by Group II (15.8 per cent) and finally Group I (25 per cent).

To test for the significance between the success rates of the three groups, and also to allow for the possible effects of the different aetiologies of the curves, the proportions were fitted to a linear logistic curve using the method of maximum likelihood (Cox 1970). It was found that the improved incidence of solid fusions in Group III compared with Groups I and II was very highly significant. Although the success rate in Group II was greater than that in Group I this was not significantly better. The success rate in each group was independent of the aetiology of the curves.

Stability of the solid fusions. Having accurately diagnosed those patients with a pseudarthrosis it was then possible to study radiographically the remaining 332 patients whose spines were known to be solidly fused. These patients were followed for periods varying from 18 months to 15 years (mean 3 years 10 months). The types of curvature are described by the area of the spine in which the apex of the curve is located (Terminology Committee of the Scoliosis Research Society 1976).

The mean loss of correction in the solidly fused curves during the first year and at final follow-up for thoracic, thoracolumbar and lumbar curves are shown in Tables III, IV and V. The correction lost was calculated

from the difference between the curvature shown on the supine spinal radiograph taken in the recovery room immediately after operation and that on the erect spinal radiograph taken at the time of follow-up. All curves were measured using the Cobb technique.

Group I curves, which were followed for an average of 4 years 10 months, had the greatest loss of correction:

after the initial loss in the first year thoracic curves subsequently lost a mean of 6.4 degrees, thoracolumbar six degrees and lumbar eight degrees. These curves also took the longest time to stabilise.

Group II curves, which were followed for an average of three years two months, had a smaller loss of correction than Group I curves before becoming stable:

Table III. Loss of correction in solidly fused thoracic scoliosis (285 curves)

	Number of curves	Mean curve before operation (degrees)	Mean correction obtained		Mean loss of correction (degrees)		
			Degrees	Per cent	At 6 months	At 1 year	Final follow-up
Group I	149	60	23	39	4.0	7.6	14.0
Group II	42	84	36	46	5.0	6.2	9.3
Group III	94	70	34	51	2.4	3.1	3.9

Table IV. Loss of correction in solidly fused thoracolumbar scoliosis (49 curves)

	Number of curves	Mean curve before operation (degrees)	Mean correction obtained		Mean loss of correction (degrees)		
			Degrees	Per cent	At 6 months	At 1 year	Final follow-up
Group I	33	68	29	47	5.0	8.0	14.0
Group II	7	72	39	59	7.7	10.8	13.5
Group III	9	89	55	64	2.5	3.2	5.3

Table V. Loss of correction in solidly fused lumbar scoliosis (20 curves)

	Number of curves	Mean curve before operation (degrees)	Mean correction obtained		Mean loss of correction (degrees)		
			Degrees	Per cent	At 6 months	At 1 year	Final follow-up
Group I	6	78	26	35	3.0	6.0	14.0
Group II	0	—	—	—	—	—	—
Group III	14	75	41	55	3.9	4.7	5.6

Table VI. Findings at exploration of 406 posterior spinal fusions

Aetiology of scoliosis	Group I		Group II		Group III	
	Number of patients	Patients with pseudarthroses (per cent)	Number of patients	Patients with pseudarthroses (per cent)	Number of patients	Patients with pseudarthroses (per cent)
Idiopathic	133	37 (27.8)	33	6 (18.1)	73	2 (2.7)
Congenital	64	13 (20.3)	9	0 (0)	25	2 (8.0)
Neuropathic	46	11 (23.9)	15	3 (20.0)	8	0 (0)
Total	243	61 (25.0)	57	9 (15.8)	106	4 (3.8)

after the losses of the first year thoracic curves lost a mean of 3.1 degrees and thoracolumbar curves 2.7 degrees. There were no lumbar curves.

Group III curves, which were followed for an average of two years two months, had the least loss of correction and all stabilised rapidly with no loss after 18 months. The mean loss of correction after the first year was no more than one degree in thoracic and lumbar curves; thoracolumbar curves lost just over two degrees.

DISCUSSION

Factors influencing the incidence of pseudarthroses

Method of correction. Harrington instrumentation is said to have helped lower the incidence of pseudarthroses by providing increased rigidity to the developing fusion. In nearly all series the pseudarthroses have been diagnosed radiographically but unfortunately radiographs of the deformed spine are often difficult to interpret. The presence of a Harrington rod may also prevent the rapid loss of correction which is often the only indication of a pseudarthrosis and this may remain unrecognised until the rod breaks due to continuing stress. In order to overcome these difficulties all of the spines in this investigation were surgically explored six months after the attempted fusion.

Group I and II curves were both fused using only the Moe (1958) interfacetal fusion technique but the method of correction was different (Table I). The incidence of pseudarthroses in Group I, where the curves were corrected in a Risser localiser jacket and the patient was kept in bed for one to three months, was 25 per cent compared with 15.8 per cent in Group II, where the patient was treated by Harrington instrumentation and mobilised in an underarm jacket at two weeks.

Moe (1958) found a 23 per cent incidence of pseudarthroses, diagnosed radiographically, in 130 patients with idiopathic scoliosis treated in a similar manner to the patients in Group I of my series. He later reported on a further 173 patients treated by Harrington instrumentation with a reduction in the incidence of pseudarthroses to 17 per cent (Moe and Valuska 1968).

Fusion technique. Harrington instrumentation and an underarm jacket were used in both Groups II and III but the technique of fusion was different (Table I). An interfacetal fusion was performed in each group, but the 106 patients in Group III had an additional total decortication of all the posterior spinal structures followed by an onlay of matchstick grafts of autogenous iliac bone. This resulted in a further significant decrease in the incidence of pseudarthroses from 16 per cent in Group II to 3.8 per cent in Group III.

Goldstein (1969) stated that total decortication followed by the addition of fresh autogenous cancellous bone grafts not only gave a lower incidence of pseudarthroses but also produced a fusion mass better

able to withstand bending once all external support was removed. In his 80 patients with idiopathic scoliosis, the incidence of pseudarthrosis, diagnosed radiographically, was five per cent. The 73 patients with idiopathic scoliosis in Group III of my series were submitted to a very similar procedure to Goldstein's except that a more meticulous technique of interfacetal fusion was used, and at exploration the incidence of pseudarthrosis was three per cent. Erwin, Dickson and Harrington (1976) compared two large series of patients with idiopathic scoliosis corrected by Harrington instrumentation and treated with and without supplementary autogenous iliac bone grafts. They reported that the incidence of pseudarthrosis, diagnosed radiographically, was reduced from 4.3 per cent to 0.6 per cent by the addition of the supplementary bone.

Homogenous and heterogenous bone grafts have also been used in other series (Blount *et al.* 1958; Brown 1965; Donaldson *et al.* 1969; Dickson and Harrington 1973) but the results have been much less successful than with autogenous bone.

Factors influencing the stability of the solidly fused spine

Postoperative management should aim to produce a solid fusion and prevent loss of correction while the fusion is maturing and gaining strength.

Bed rest. Opinion regarding the appropriate period of bed rest has varied considerably. From 1935 to 1964 Cobb kept all his patients with scoliosis in bed for nine months after operation—six months in a Risser turnbuckle jacket, followed by three months in an underarm jacket. In 1958 Risser and Norquist reported that walking in a Risser localiser plaster jacket was possible 7 to 10 days after operation without significant loss of correction or increase in the incidence of pseudarthrosis. Most surgeons, however, continued to keep their patients in bed for at least three months and Wilson, Levine and Doherty (1971) extended this period to six months, without improving their results.

Clinical experience and experimental studies have shown that early weight-bearing not only accelerates the rate of healing of fractures but also significantly improves the strength of the healing bone (Sarmiento *et al.* 1977). Theoretically the same principles should apply to the spine after fusion and the early transmission of weight-bearing forces should strengthen and increase the size of the fusion mass. The psychological advantages of decreasing the period of recumbency and the time in hospital are obvious. In 1973 Leider, Moe and Winter reported that since 1967 they had allowed their patients to walk in a Risser-Cotrel jacket 7 to 15 days after Harrington instrumentation with only slight loss in correction; in their opinion the type of cast used was the key to success. Erwin *et al.* (1976), however, considered the full Risser cast unnecessary and since 1970 have allowed their patients to walk 7 to 10 days after

Harrington instrumentation in a simple underarm body cast, with even less loss of correction and a very low incidence of pseudarthroses.

This theory of early mobilisation was put into practice in the management of the patients in Group III who were allowed to walk 7 to 10 days after Harrington instrumentation in a well-fitting underarm jacket and returned to normal activities. In contrast, the patients in Group I were kept in bed in a localiser jacket for one to three months after which their activities were confined to the house and garden until six months after operation. The patients in Group II were treated in an intermediate manner; they were confined to bed for two to four weeks after Harrington instrumentation and then allowed home in an underarm plaster jacket and confined to the house and garden until six months after operation (Table II). Group III curves which were treated by early mobilisation had better maintenance of correction than the curves in Group II which were better than the curves in Group I (Tables III to V).

Type and duration of immobilisation in plaster. The time spent in a plaster jacket after spinal fusion has also varied. Ponseti and Friedman (1950) found that the rate of pseudarthrosis was not influenced by cast immobilisation for periods longer than five months. Erwin *et al.* (1976) found, however, that if the plaster jacket was removed at six months there was an increased loss of correction up to 12 months with only a minimal loss occurring subsequently. This loss of correction in an otherwise solid fusion was presumably due to the immaturity of the fusion and prompted them to extend the length of time in plaster to nine months.

The type and duration of external spinal support for the solidly fused curves in my series varied in each of the three groups (Table II). The greatest loss of correction in the first year occurred in Group I where the spine was supported by a Risser localiser jacket for six months followed by six months in a Milwaukee brace. Harrington instrumentation and an underarm jacket were used in both Groups II and III, but the time in plaster and the fusion technique were different. In Group II the jacket was maintained for one year whereas for the majority of patients in Group III it was removed after ten months. Despite this Group III curves maintained their correction better and this can only be attributed to a greater

strength in the fusion due to the use of the supplementary bone grafts.

Fusion technique. External support was not used after the first year for any of the solidly fused curves. These curves then suffered further loss of correction due to bending of the immature fusion before finally becoming stable. The severity of this loss was different in each of the three groups but the majority of curves had stabilised by 18 months. Although the same fusion technique was used in Groups I and II, there was a better maintenance of correction in Group II curves and this can only be explained by the presence of the Harrington internal fixation. Harrington instrumentation was used in both Groups II and III but the fusion technique was different. Group III curves maintained their correction better, which can be attributed to a stronger and more rapidly maturing fusion.

The total loss of correction in Group III by the time the spines had stabilised was a mean of 3.9 degrees for thoracic, 5.3 degrees for thoracolumbar and 5.6 degrees for lumbar curves. This loss of correction is similar to that in two other series treated by Harrington instrumentation with supplementary autogenous iliac bone grafts and early mobilisation. Leider *et al.* (1973) studied 106 patients with idiopathic scoliosis followed for an average of 19 months with an overall loss of correction of five degrees. In the series of 177 patients with idiopathic scoliosis, followed for an average of two years by Erwin *et al.* (1976), the overall loss of correction was 4.3 degrees.

My conclusions therefore are as follows. Early mobilisation 7 to 10 days after Harrington instrumentation, and a return to normal activities in a well-moulded underarm plaster jacket, does not have a detrimental effect on the maintenance of correction and may possibly have a beneficial effect on the development of the fusion. Total decortication and an onlay of autogenous iliac bone grafts in addition to a Moe interfacetal fusion and Harrington instrumentation not only lowers the incidence of pseudarthroses but also creates a spinal fusion which rapidly becomes stable with only minimal loss of correction after removal of all external support at 10 months. With these more modern techniques routine diagnostic exploration of the spine is no longer necessary.

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Pseudarthrosis: A Late Cause of Paraparesis after Scoliosis Surgery

A CASE REPORT

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Pseudarthrosis and fracture of a Harrington rod are recognized complications of the treatment of scoliosis by posterior spine fusion. To our knowledge, there is no record of a pseudarthrosis causing spinal-cord compression as a late complication after posterior fusion for scoliosis. We are reporting such a case.

Case Report

An obese fifty-nine-year-old woman was seen by one of us (M. J. McM.) in August 1976, complaining of backache that had become increasingly severe over a number of years. Radiographs of the spine showed a severe idiopathic left lumbar scoliosis extending from the twelfth thoracic to the fourth lumbar vertebra and measuring 84 degrees. In addition, there were severe degenerative changes in the spine in the region of the curvature as well as two stress fractures of the twelfth rib, which abutted against the right iliac crest (Fig. 1). Movement of the lumbar spine was limited and painful, but neurological examination revealed normal sensation and motor power in both lower limbs. Straight-leg raising was pain-free and of normal range on both sides. Deep tendon reflexes were present and equal bilaterally, and both plantar responses were flexor.

To rule out spinal stenosis, a preoperative myelogram was performed. It showed no evidence of cord compression or stenosis.

At operation in November 1976, the scoliosis was partially corrected using a Harrington distraction rod, and a posterior spine fusion was performed from the twelfth thoracic vertebra to the sacrum. The joints between the articular facets were fused using the Moe technique, followed by decortication of the lateral masses and transverse processes, and onlay corticocancellous iliac-bone grafts were placed in both paraspinal gutters.

The postoperative course was uneventful. Radiographs showed that the curve had decreased to 56 degrees and the twelfth rib no longer abutted against the iliac crest (Fig. 2). There were no postoperative neural abnormalities. One week after surgery an underarm plaster jacket was applied and the patient was allowed to walk.

After nine months, at which time the plaster jacket was removed, the patient was free of pain and radiographs showed an apparently solid fusion. No further external support was thought to be necessary. The patient was followed clinically and radiographically without apparent problems for another six months. She remained asymptomatic until January 1980 when she had sudden, severe back pain followed by gradually increasing weakness in both lower limbs, which after three weeks prevented her from walking. On examination, there was generalized weakness in both lower limbs and the strength of all muscle groups was less than 5. The most severe weakness was on the right side, where the hip flexors and quadriceps were rated 3 and the ankle extensors and evertors were completely paralyzed. The ankle and knee jerks were hyperactive bilaterally. The plantar response was extensor on the right and equivocal on the left. Sensory, bladder, and bowel function was normal. A radiograph of the spine showed a fracture of the Harrington rod at the junction

of the smooth and ratcheted portions, a pseudarthrosis in the fusion mass between the twelfth thoracic and first lumbar vertebrae, and a 4-degree loss of correction (Fig. 3). A myelogram was performed using a cisternal puncture, and this revealed a partial obstruction due to a posterior indentation at the level of the pseudarthrosis (Fig. 4).

The spine was immediately explored posteriorly through the original incision. The normal architecture of the spine was obscured by the fusion but a pseudarthrosis composed of an overgrowth of soft bone and fibrous tissue could be seen running transversely across the fusion mass, through the interlaminar space between the twelfth thoracic and first lumbar vertebrae, and bilaterally into the regions of the joints between the articular facets. Starting at the midline, the pseudarthrosis was excised using fine osteotomes and bone rongeurs. The underlying dura was compressed by soft bone and fibrous tissue and did not pulsate until the laminae of the twelfth thoracic and first lumbar vertebrae had been removed and the pseudarthrosis was excised bilaterally into the regions of the joints between the facets. The remainder of the fusion was seen to be solid and the broken Harrington rod was removed. The spine was then stabilized at the site of the pseudarthrosis using two Harrington compression rods, and strips of corticocancellous bone from the iliac crest were placed bilaterally, leaving the dura exposed.

Postoperatively, the patient regained neural function and was able to walk with crutches at three weeks. After three months there were no neural abnormalities. Radiographs of the spine made nine months after decompression (Fig. 5) showed that the fusion was solid on both sides at the site of the pseudarthrosis and there was no increase in the curve. At the time of writing the patient had been followed for two years and remained free of pain, with normal neurological findings.

Discussion

The association of a pseudarthrosis after spine fusion for scoliosis and the late onset of neural abnormalities has not been described previously. The incidence of neural complications following scoliosis surgery was reported by the Scoliosis Research Society to be 0.72 per cent⁴. These complications were all detected in the immediate postoperative period and were due to either direct injury to the spinal cord at the time of surgery or excessive traction on the cord while attempting to correct the deformity. The only previously reported case of late onset of neural symptoms was in a nineteen-year-old man with an adolescent idiopathic thoracic scoliosis, two years after posterior spine fusion². At surgical exploration, the spinal cord was found to be compressed by a localized overgrowth of bone from the fusion mass, but there was no evidence of a pseudarthrosis.

Neural symptoms developed in our patient more than three years after posterior spine fusion and were due to compression of the spinal cord by an overgrowth of bone and fibrous tissue at the site of a pseudarthrosis. The sud-

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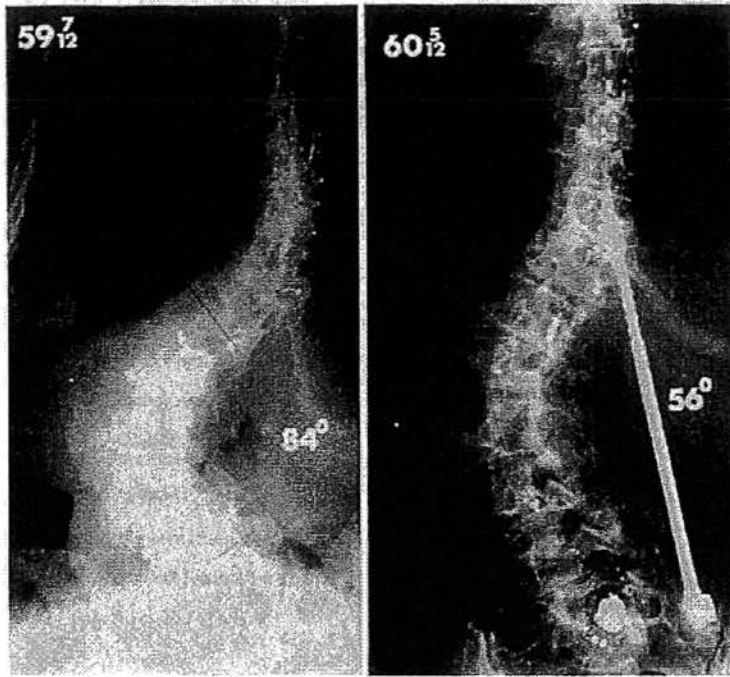


FIG. 1

FIG. 2

Fig. 1: Initial radiograph showing a left lumbar scoliosis, degenerative changes in the lumbar spine, and stress fractures of the right twelfth rib caused by impingement against the iliac crest.

Fig. 2: Postoperative radiograph showing the amount of correction obtained.

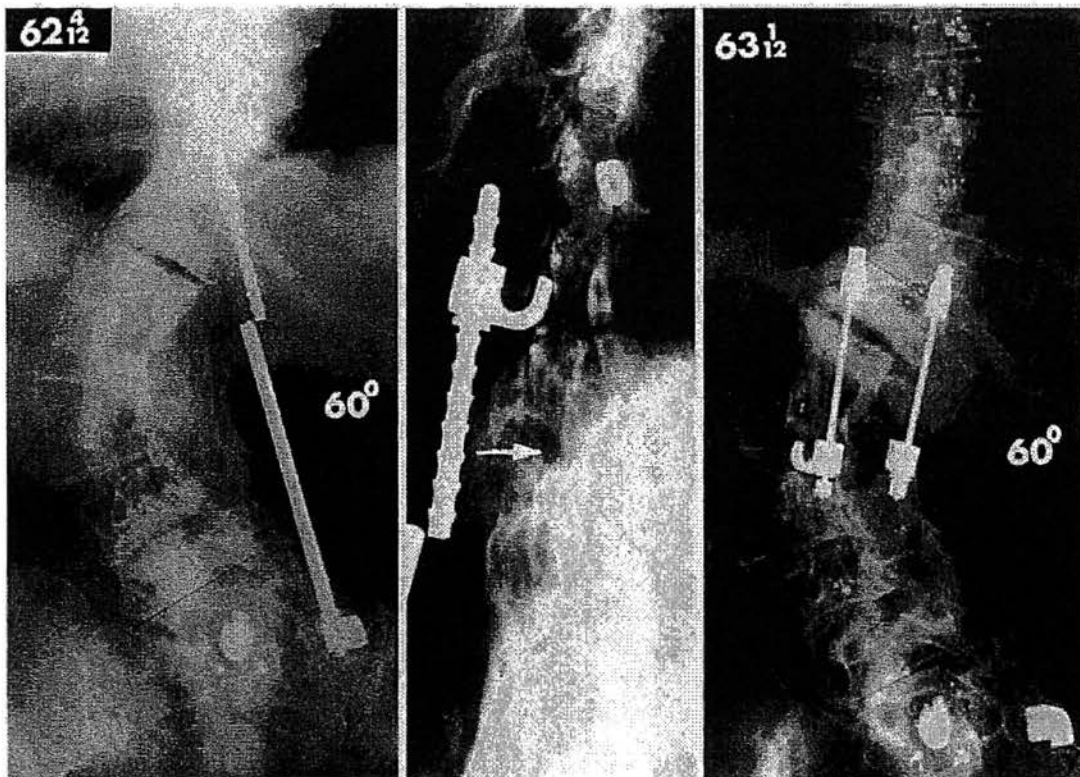


FIG. 3

FIG. 4

FIG. 5

Fig. 3: More than three years after fusion there is a fracture of the Harrington rod, an obvious pseudarthrosis between the twelfth thoracic and first lumbar vertebrae, and a 4-degree loss of correction.

Fig. 4: Myelogram showing posterior indentation of the column of contrast medium (arrow) at the site of the pseudarthrosis.

Fig. 5: Radiograph made nine months after the second fusion, showing the pseudarthrosis stabilized by Harrington compression rods and a solid fusion.

den onset of back pain was most likely due to the fracture of the Harrington distraction rod which had previously splinted the pseudarthrosis. Slight displacement of the twelfth thoracic vertebra at the pseudarthrosis after fracture of the rod probably was the final precipitating factor in the development of the spinal cord compression. The reported incidence of pseudarthrosis after posterior spine fusion for scoliosis in adults has ranged from 10 to 27 per cent^{1,3,6,7}. It is therefore surprising that spinal cord com-

pression is not more common, especially in older patients who may already have an element of spinal stenosis due to degenerative changes. In our patient, resection of the pseudarthrosis and adequate stabilization resulted in a painless spine and complete return of normal neural function over a three-month period. The surgeon must be aware of this late complication after an apparently successful early result and must follow patients carefully after fusion for scoliosis.

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The surgical management of adolescent idiopathic scoliosis in Edinburgh 1975–1982

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The treatment of scoliosis has shown significant advances in preoperative, operative and postoperative management in the last two decades. The surgical management of 120 consecutive patients with adolescent idiopathic scoliosis at the Edinburgh Scoliosis Clinic between 1975 and 1982 is discussed.

Adolescent idiopathic scoliosis is a structural curve which develops in healthy teenagers for no apparent reason and which frequently continues to progress until skeletal maturity. The smaller curves can be satisfactorily treated by conservative means but larger curves require surgery, not only to correct the deformity but also to prevent further deterioration. Harrington instrumentation has been the method most frequently used to correct the scoliosis as much as possible. Following correction a posterior spinal fusion is performed whose success depends on transforming the structural curve into a solid bar of bone which is of sufficient strength to maintain the correction and resist bending under the continuing influence of the factors which caused the deformity. Correction will be lost if the fusion is weak or a pseudarthrosis develops.

The Edinburgh Scoliosis Clinic is a referral centre for patients with adolescent idiopathic scoliosis from Scotland and elsewhere (Fig 1). The care of these patients before and after surgery varies considerably between centres and in this paper we report on the surgical management in Edinburgh between 1975 and 1982 with special emphasis on a short hospital stay followed by an early return to school and normal activity in a simple underarm plaster jacket.

Clinical material

At the Princess Margaret Rose Orthopaedic Hospital, Edinburgh, during the period 1975–1982 120 consecutive patients with adolescent idiopathic scoliosis were treated by Harrington instrumentation and a posterior spinal fusion. All the operations were performed by one of the authors (M J McM). The same operative technique and after-care were employed in all cases. There were 94 girls and 26 boys. The mean age at operation was 14 years 6 months (range 10 years 2 months to 18 years 10 months).

91 patients had a single thoracic curve and 7 patients a single thoracolumbar curve. 3 patients had double thoracic curves and 19 patients double curves affecting the thoracic and lumbar regions. 110 of the curves were to the right and 31 to the left. The mean preoperative curve and range for each group is shown in Table 1.

After surgery the patients were followed from a minimum of one year 3 months to 4 years 8 months (mean 2 years 1 month). 87 patients were followed to skeletal maturity.



Fig 1 Geographical distribution of adolescent idiopathic scoliosis patients treated surgically at the Edinburgh Scoliosis Unit.

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Methods

Operative technique. The scoliosis was corrected at the time of surgery using a single distraction rod but without the compression system¹. There was no preoperative

TABLE 1
Operative treatment of adolescent idiopathic scoliosis
(120 patients)

Type of curve	No of curves		Mean curve preoperative* (degrees)	Mean curve correction postoperative*		Loss of correction at final follow-up* (degrees)
	Right	Left		Degrees*	Percentage*	
Thoracic	83	8	62 (30-122)	34 (5-59)	55 (16-80)	4 (1-13)
Thoracolumbar	7		52 (36-75)	34 (20-47)	65 (48-82)	10 (4-16)
Double thoracic		3	42 (26-50)	16 (12-15)	39 (30-46)	3 (1-5)
thoracic	3		54 (36-67)	23 (14-36)	46 (39-54)	5 (1-8)
Double thoracic	19		60 (40-85)	29 (16-45)	47 (28-70)	4 (2-15)
lumbar		19	57 (36-83)	31 (22-47)	54 (36-74)	6 (2-17)

*Numbers in parentheses are ranges.

traction or the use of corrective plaster jackets. The spine was fused posteriorly from the upper to the lower Harrington hooks by the interfacetal, intertransverse fusion technique described by Moe². There was then a deep and thorough decortication of all the posterior bony structures from the midline out to the tips of the transverse processes. This was followed by the application throughout the fusion area of large amounts of fresh autogenous iliac bone grafts which had been cut into matchsticks. A mean of 10 vertebrae (range 8-13 vertebrae) were fused in single curves and a mean of 13 vertebrae (range 11-15 vertebrae) in double curves.

Postoperative care. After operation all patients were nursed free in bed for 7-10 days and a simple well moulded underarm plaster jacket was then applied with the patient lying without traction on the spinal table. The patients were mobilised, and allowed home within a few days. They returned to school and normal activities in 2 to 3 weeks, the only restriction being contact sports. The jacket (Fig 2) was worn for 9 to 10 months and was changed because of growth, hygiene requirements or general deterioration, on average once in this time. The more recent jackets, constructed from the newer casting materials such as Baycast, have proved to be much lighter, more hygienic and durable but require greater experience to apply. When the underarm jacket was finally removed the presence of pseudarthroses was excluded radiographically. The patients were then allowed unrestricted activities without external support.

Results

The severity of the scoliosis was measured using the Cobb method³ on the erect anteroposterior spinal radiographs taken before operation, immediately after surgery and at final follow-up.

Degree of correction. The number of degrees of correc-



Fig 2 Underarm jacket. It is fitted 7-10 days after surgery and worn for 8 months.

tion and the percentage correction compared with the original curve for each site of the scoliosis are shown in Table 1. The mean correction in the single thoracic curves was 34° (55%) and in the thoracolumbar curves 34° (65%). In the double thoracic curves the upper curve was corrected by a mean of 16° (39%) and the lower curve by a mean of 23° (46%). In the double curves affecting the thoracic and lumbar regions the thoracic curve was corrected by a mean of 29° (47%) and the lumbar curve by a mean of 31° (54%).

Loss of correction. The mean loss of correction between the radiograph taken immediately after surgery and at

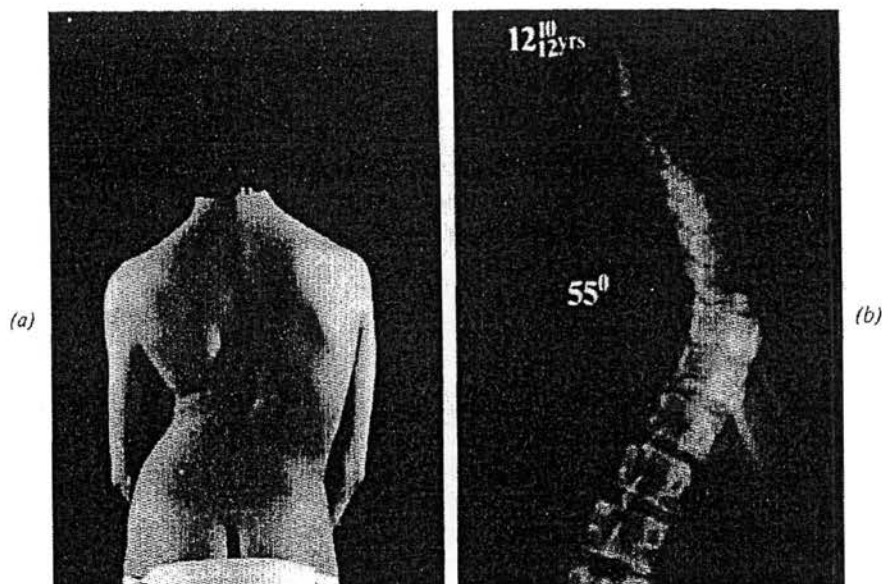


Fig 3 • Preoperative photograph (a) and radiograph (b) of a girl aged 12 years 10 months with a thoracic curve of 55° (measured by Cobb method).

final follow-up for each of the different sites of curve are shown in Table 1. The least loss occurred in single thoracic curves (Figs 3–5) and was a mean of 4° (range 1–13°). The greatest loss occurred in thoracolumbar curves and was a mean of 10° (range 4–16°). The mean loss of correction for all of the curves was 5°.

Complications. Immediately following surgery 2 patients complained of paraesthesia affecting the soles of their feet but this recovered completely in 24–48 hours. Another patient complained of paraesthesia and slight weakness affecting one hand. This was thought to be due to a mild stretch of the lower roots of the brachial plexus caused by hyperabducting the arm when the patient was prone during the operation. The paraesthesia and weakness ceased after a few weeks.

3 patients developed superficial infections of the iliac crest wound through which the bone graft had been taken. These responded rapidly to local care and antibiotic therapy. 2 patients developed minor wound separations which required resuturing; satisfactory healing ensued.

Chest infections from postoperative atelectasis occurred in 2 patients and rapidly responded to antibiotics and physiotherapy.

Shortly after the application of the plaster jacket 2 patients developed paraesthesia in the distribution of the lateral cutaneous nerve of the thigh. This recovered completely when the plaster was trimmed.

A pseudarthrosis in the fusion developed in one patient (0.8%)—a 13-year-old girl with a thoracolumbar scoliosis which had been corrected from 45° to 8°. 6 months after surgery there was only 9° loss of correction but a possible pseudarthrosis was noted radiographically; this was confirmed at surgical explora-

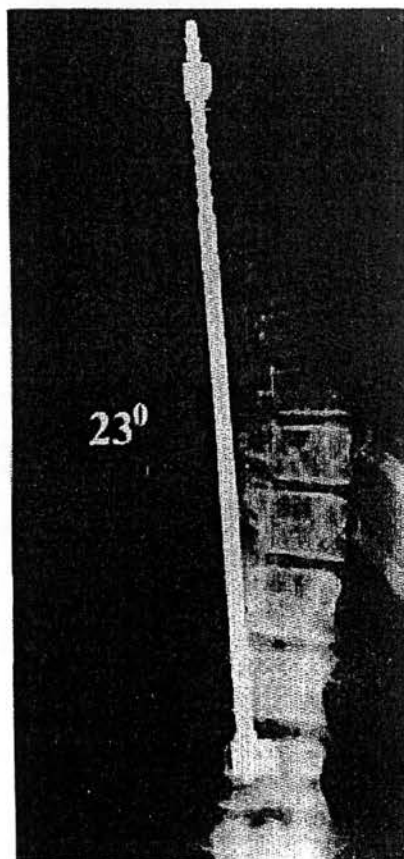


Fig 4 Postoperative radiograph of the same patient showing correction of the curve from 55° to 23°. The position of the Harrington rod is shown.

ation and repaired. After a further 5 months in plaster the spine became solidly fused. The patient has been followed to skeletal maturity with only 7° loss of correction.

There were no complications due to failure of the Harrington instrumentation or slipping of the hooks.

Duration of hospital stay. The mean duration of hospital stay was reduced from 24 days in the first 2 years of the series to a mean of 18 days (range 11–45 days) in the last 5 years. All patients returned to school 2–3 weeks after

posterior spinal structures and an onlay of autogenous bone grafts⁴.

Erwin *et al*⁷ compared two groups of patients with adolescent idiopathic scoliosis. Both groups were corrected by Harrington instrumentation and treated by an interfacetal fusion but in one group there was a supplementary onlay of autogenous iliac bone grafts. The incidence of pseudarthrosis was reduced from 4.3% to 0.6% by the addition of the supplementary bone. The incidence of pseudarthrosis in our series was 0.8% (one

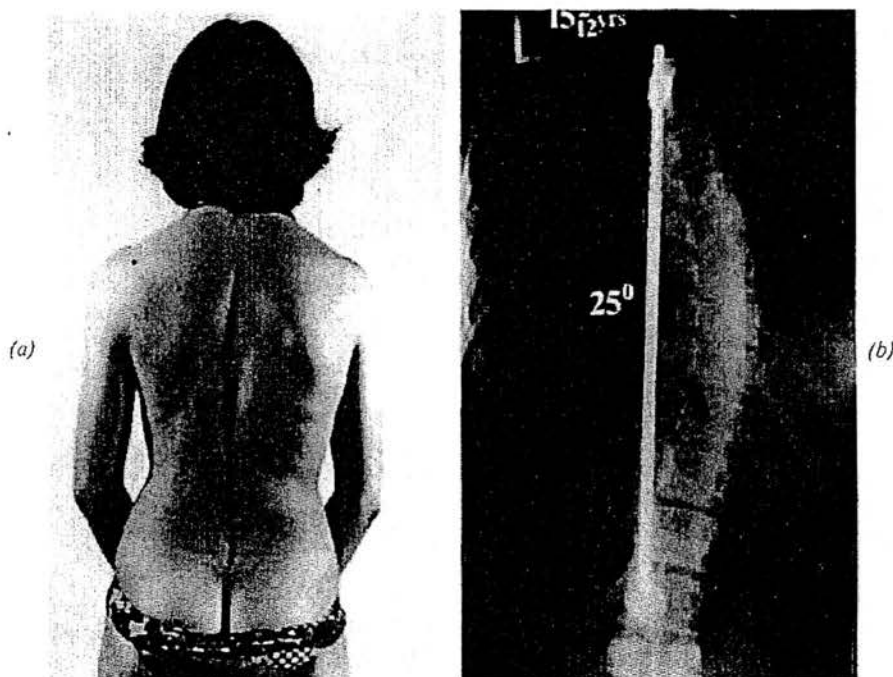


Fig 5 (a) Appearance at skeletal maturity.
(b) Same patient. Radiograph at skeletal maturity showing loss of 2° since operation.

discharge from hospital and none complained of problems with their schooling. After the plaster jacket was removed the patients returned to normal activities without difficulty.

Discussion

Since the introduction of Harrington instrumentation¹ and modern fusion techniques^{2,4} there has been a significant improvement in the treatment of adolescent idiopathic scoliosis. Greater degrees of correction of the curvature can be obtained and a solid fusion is more predictable. Moe and Valuska⁶ found that with Harrington instrumentation, the incidence of pseudarthrosis in idiopathic scoliosis was reduced from 23% to 17%. Harrington instrumentation provides increased rigidity and is therefore thought to encourage the development of a solid fusion. The incidence of pseudarthrosis has also been further reduced by fusion techniques which combine an interfacetal fusion with decortication of the

patient) and we attribute the very low incidence to the use of Harrington instrumentation and a fusion technique which combines a meticulous interfacetal, intertransverse fusion followed by total decortication of all the posterior bony elements and a massive onlay of fresh autogenous iliac bone grafts.

The risk of neurological complications following the operative correction of scoliosis has been estimated to be 0.2%⁵. It has been argued that preoperative traction techniques using Cotrel traction would lead to improved operative correction with less risk of neurological complications⁸. In our series preoperative traction was not used and the mean degree of correction was 55% for thoracic curves and 65% for thoracolumbar curves, figures which compare favourably with the other series^{4,7,9}, and there were no neurological complications attributable to the intraoperative correction. In our opinion preoperative traction unnecessarily prolongs the stay in hospital and others have found that the correction

obtained is no better than that normally obtained at surgery by Harrington instrumentation alone¹⁰.

The aim of postoperative management following scoliosis surgery is to encourage the development of a solid fusion and to prevent loss of correction while the fusion is maturing and gaining strength¹¹. Opinions differ as to the best methods of achieving this. Until 1964 Cobb kept all his scoliosis patients in bed for 9 months after operation. In 1958 Risser and Norquist¹² reported that walking in a localiser plaster jacket, which extended to incorporate the neck, was possible 7–10 days after operation without significant loss in correction. However, despite this report, most surgeons continued to keep their patients in bed for up to 3 months. In 1973 Leider *et al*⁹ reported that following Harrington instrumentation they mobilised their patients in a Risser jacket incorporating the neck, 7–10 days after operation with only a slight loss of correction. Erwin *et al*⁷, however, considered the full Risser jacket unnecessary and allowed early mobilisation in a simple underarm jacket. A similar policy has been used in Edinburgh since 1975 and patients have been allowed to walk in a lightweight underarm jacket 7–10 days after surgery and return to school and normal activities, with the exception of contact sports, within one month of operation. The psychological advantages of this regime have proved to be considerable.

The length of time that the plaster jacket should be worn remains controversial¹³. Erwin *et al*⁷ found that if it was removed at 6 months there was an increased loss of correction over the following 6 months with only minimal loss occurring subsequently. This loss of correc-

tion in an otherwise solid fusion was presumably due to plasticity of the immature bone and prompted them to extend the length of time in plaster to 9 months. We have maintained the plaster jacket on our patients for 9 months and found that the mean loss of correction at final follow-up when the spine had stabilised was 4° for thoracic curves and 10° for thoracolumbar curves. The overall mean loss was 5°. This compares favourably with other series^{4,7,9}. Leider *et al*⁹ studied 106 patients with idiopathic scoliosis followed for an average of 19 months with an overall mean loss of correction of 5°. In a series of 177 patients with idiopathic scoliosis followed for an average of 2 years, Erwin *et al*⁷ found the overall mean loss of correction to be 4.3°. More recently we have reduced the period in plaster to 8 months without apparent increase in the loss of correction and consider this to be attributable to the strength of the fusion produced by the large onlay of autogenous bone grafts.

In conclusion the operative treatment of idiopathic scoliosis has radically improved over the last two decades. It is no longer necessary for these children to be in hospital for long periods and in Edinburgh the hospital stay has been reduced to an average of 18 days. An early return to school and normal activities in a simple underarm jacket is possible with very little inconvenience and without detriment to the result of their treatment. The next stage in the evolution of the surgical treatment of idiopathic scoliosis is to provide a method of internal fixation which is sufficiently stable to allow unrestricted activities without external immobilisation and still produce results as good and as safe as those with Harrington instrumentation.

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THE USE OF INDUCED HYPOTENSION TO CONTROL BLEEDING DURING POSTERIOR FUSION FOR SCOLIOSIS

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The operative and anaesthetic technique for 44 patients undergoing posterior spinal fusion with Harrington rod instrumentation for idiopathic scoliosis is described. There were two groups of 21 and 23 patients, matched for diagnosis and status before operation. The management of both groups was similar but in one group anaesthesia with induced hypotension was employed, using a mixture of sodium nitroprusside and trimetaphan. The mean blood loss at operation and after operation in this group was significantly lower than in the other group, with a consequent reduction in the transfusion requirement. No adverse sequelae were observed. All patients showed a drop in haemoglobin concentration after operation, despite clinically adequate blood transfusion.

Procedures for surgical correction and posterior spinal fusion of patients with scoliosis are associated with considerable haemorrhage during and after operation. Induced hypotension has been established as a means of reducing bleeding during such operations (McNeill *et al.* 1974; Bennett and Abbott 1977; Grundy, Nash and Brown 1979; Abbott and Bentley 1980) although some authors have questioned its safety (Relton 1977). However, previous reports comparing groups of patients undergoing normotensive and hypotensive anaesthesia have failed to match the two groups: the aetiology of the scoliosis has varied, the operations have been carried out by a number of surgeons and the technique of operation has not always been the same.

The purpose of this paper is to compare the blood loss during and after operation in two groups of patients who were operated on by the same surgeon and accurately matched for age and size and for the aetiology of their scoliosis. The only difference was that in one group the patients were studied prospectively and had their blood pressure deliberately lowered below that occurring as a normal consequence of the anaesthetic technique. Hypotension was induced by a modification of the method employed by MacRae, Wildsmith and Dale (1981), using a mixture of sodium nitroprusside and trimetaphan. In their study the technique combined the advantage of ease of control of blood pressure with a low dose of the potentially toxic sodium nitroprusside.

MATERIAL AND METHODS

A study was made of two groups of male and female patients with idiopathic scoliosis who underwent posterior spinal fusion with Harrington rod correction at the Princess Margaret Rose Orthopaedic Hospital. The patients were aged between 10 and 18 years with an average age of 14.4 years, and all weighed over 45 kilograms. The anaesthetic technique was similar in all patients except that induced hypotension was employed in 23 patients (Group II) while 21 patients (Group I) were kept normotensive. The general health of all of the patients was good, with no significant difference in the mean systolic blood pressure or haemoglobin concentration between the two groups. All of the operations were performed by one of us (MJMcM) using an identical technique. An average of 10.8 vertebrae were fused in Group I and 10.1 in Group II (no significant difference at the five per cent level). The operations in Group I took place between 1977 and 1979 and in Group II between 1980 and 1982.

Anaesthetic technique. Induction of anaesthesia was by an intravenous dose of thiopentone, methohexitone or Althesin to produce sleep, followed by a non-depolarising muscle relaxant (alcuronium 0.2 milligrams per kilogram of body weight, pancuronium 0.1 milligrams per kilogram of body weight or *d*-tubocurarine 0.4 milligrams per kilogram of body weight), and an opiate analgesic (phenoperidine 0.5 to 1.5 milligrams or fentanyl 0.05 to 0.15 milligrams). The patient was intubated and intermittent positive-pressure ventilation established with a Cape Waine ventilator set to a minute volume of 120 millilitres per kilogram of body weight. Anaesthesia was continued with nitrous oxide and one per cent halothane, using an inspired oxygen concentration of at least 30 per cent rising to 50 per cent during the period of induced hypotension. Incremental doses of analgesic were given

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if indicated by the development of tachycardia, and muscle relaxant was given if venous bleeding became obvious. Blood pressure was measured in Group I with an oscillotonometer or with a sphygmomanometer and peripheral-pulse meter. In Group II direct intra-arterial measurement with a Bentley Trantec transducer and Roche 224-1 monitor was used. Throughout the operation there was continuous electrocardiographic monitoring.

Fusion technique. The position of the patient on the operating table, the surgical approach, the method of correcting the scoliosis and the fusion technique were identical in both groups of patients.

The patient was positioned prone on a specially designed foam mattress in which a hole had been cut allowing the entire abdomen to hang free of the table and mattress. The table was tilted so that the patient's head was approximately 10 degrees head down, and broken to allow the patient's hips to be flexed. By this method the intra-abdominal pressure was minimised and venous bleeding decreased.

A straight midline incision was made over the spine after infiltrating the intradermal and subcutaneous tissues with up to 40 millilitres of adrenaline solution (1:500 000) to reduce capillary bleeding. The soft tissues were stripped subperiosteally, starting over the tips of the spinous processes and extending laterally out to the tips of the transverse processes. Using this technique the whole of the scoliosis was exposed and the bleeding points controlled with diathermy.

Correction of the scoliosis was obtained by means of Harrington instrumentation using a single distraction rod (Harrington 1962). Corticocancellous bone grafts were taken from the outer cortex of the right iliac crest. In patients with thoracolumbar or lumbar curves, the grafts were taken through the same incision but in patients with thoracic curves a second incision over the iliac crest was necessary. The gluteal muscles were stripped subperiosteally and after the grafts had been taken an area of bleeding cancellous bone approximately six centimetres by six centimetres was exposed. Bone wax was applied to the area to control bleeding and a closed suction drain inserted into the dead space beneath the muscles.

The spine was fused by excising the interfacetal joints and packing them with corticocancellous iliac bone grafts (Moe 1958). This was followed by a deep and thorough decortication of all the posterior bony structures from the midline out to the tips of the transverse processes. This exposed a large area of bleeding cancellous bone over which generous iliac onlay grafts were placed throughout the fusion area. Two closed suction drains were inserted and the wound closed in layers.

Technique of inducing hypotension. Hypotension in Group II patients was induced by an infusion of 25 milligrams of sodium nitroprusside and 125 milligrams trimetaphan in 500 millilitres of a five per cent solution of dextrose.

The infusion rate was started at 60 millilitres per hour but adjusted, according to the response of the individual patient, to reduce the systolic blood pressure to between 60 and 70 millimetres of mercury; this was equivalent in all cases to a mean arterial pressure of 50 millimetres of mercury or greater. The infusion rate usually lay between 30 and 60 millilitres per hour, though in one case 150 millilitres per hour was required. The infusion was discontinued as the wound was being closed. The blood pressure rose within five minutes of discontinuing the infusion and in no case did hypotension persist, even in those in whom the manufacturer's recommended dosage of sodium nitroprusside (1.5 micrograms per kilogram of body weight per minute) had been exceeded. The operative time was approximately three hours in all cases; the duration of induced hypotension, where it was used, was over two hours.

Fluid replacement and measurement of blood loss. An infusion of saline or Hartmann's solution was started initially and replaced by blood when the loss had reached 500 millilitres or 10 per cent of the blood volume. Thereafter blood was given in amounts equivalent to the blood loss which was assessed by the measurement of blood in the suction bottle and by weighing of swabs.

After-care. Muscular relaxation was reversed by atropine 0.02 milligrams per kilogram of body weight and neostigmine 0.04 milligrams per kilogram of body weight. After operation the patients were observed in a recovery area for six hours before being returned to the ordinary ward. The tissue drains were opened at four-hourly intervals and the blood loss recorded. Requirement for blood replacement was assessed both by the drainage loss and by the state of the circulation. On the second day after operation the drains and the intravenous infusion were removed. In all cases the circulation was stable at this time and there was no further blood loss. Blood was taken for haemoglobin estimation.

RESULTS

The systolic blood pressures for both groups before, during and after operation are shown in Table I.

The blood losses for both groups with their standard deviations are shown in Table II. The mean loss during operation in Group II was 525 millilitres compared with 1530 millilitres in Group I. The mean total loss in Group II was 1058 millilitres compared with 2544 millilitres in Group I. The mean loss after operation in Group I (1014 millilitres) was also significantly greater than that in Group II (533 millilitres). These findings are reflected in the transfusion requirements (Table III). The mean total number of units of blood required in Group I was 4.9 compared with 2.2 in Group II.

A comparable drop in haemoglobin concentration was observed in both groups after operation despite clinically adequate blood replacement (Table IV).

There were no neurological complications relating to either the brain or spinal cord in either group. All the

Table I. Systolic blood pressures before, during and after operation

Group	Number of patients	Mean systolic blood pressure (millimetres of mercury)				
		Before operation	During operation	After completion of operation (still under anaesthesia)	One hour after operation	Four hours after operation
I	21	121	87	98	122	121
II	23	117	65*	109†	117	114

*Pressures significantly lower in Group II ($P < 0.01$)†Pressures significantly higher in Group II than in Group I ($0.05 > P > 0.02$)Table II. Blood losses during and after operation (mean \pm standard deviation)

Group	Number of patients	Loss during operation (millilitres)	Loss after operation (millilitres)	Total loss (millilitres)
I	21	1530 \pm 941	1014 \pm 558	2544 \pm 1260
II	23	525 \pm 226*	533 \pm 267*	1058 \pm 339*

*Blood loss significantly less in Group II than in Group I ($P < 0.001$)Table III. Blood transfusion requirements (mean \pm standard deviation)

Group	Number of patients	Average number of units		
		During operation	After operation	Total
I	21	3.3 \pm 1.6	1.6 \pm 1.1	4.9 \pm 2.3
II	23	1.3 \pm 0.8*	0.9 \pm 0.7*	2.2 \pm 0.9*

*Amount of blood transfused was significantly less in Group II than in Group I ($P < 0.001$)Table IV. Changes in haemoglobin levels after operation (mean \pm standard deviation)

Group	Number of patients	Haemoglobin level (grams per decilitre)		Drop in haemoglobin level (grams per decilitre)
		Before operation	After operation	
I	21	13.6 \pm 0.9	10.6 \pm 2.0	-3.0 \pm 1.9
II	23	13.5 \pm 1.1	11.2 \pm 1.3	-2.3 \pm 1.4

No significant difference between the two groups

wounds healed satisfactorily and there were no infections.

The results were analysed using Student's *t* test for unpaired samples.

DISCUSSION

This study confirms that bleeding associated with posterior spinal fusion for scoliosis can be safely controlled by induced hypotension. Our patients were carefully matched for age, size, diagnosis, operation and anaesthesia. The only difference between the two groups was the induced hypotension during the operation, indicating that it was this rather than any other variable which reduced the bleeding. The reduction in mean total blood loss between the two groups was 58 per cent.

The common criticism that induced hypotension leads to increased bleeding after operation was not supported by our findings. In fact the group of patients who had undergone hypotensive anaesthesia lost less blood after operation than those who had been normotensive throughout. The reason for this is uncertain. Two factors that influence bleeding are the blood pressure and the integrity of the coagulation mechanism. In our patients the systolic blood pressures after operation in both groups were similar and no patient had clinical

evidence of a coagulopathy. Jacobs, Asher and Gilbert (1980) found in their patients with spinal fusion that a coagulopathy was present following haemorrhage and transfusion, although their comparable patients required larger volumes of donor blood (average 65 millilitres per kilogram). Therefore it appears that either a higher blood loss is associated with the blood transfusion itself, or the circulatory change leading to reduction in bleeding persists after hypotensive anaesthesia by the technique we have described.

A fall in haemoglobin after operation was found in both groups despite the apparent complete replacement of blood loss during and after operation. This change could be explained by insufficient blood replacement, by haemodilutional anaemia, or by partial haemolysis of stored blood. In no patient was the haemoglobin after operation so low that late transfusion was needed.

It has been suggested that hypotension during operations for scoliosis may lead to neurological damage due to decreased blood supply to the brain and spinal cord (Hardy *et al.* 1973). This risk is theoretically increased by the use of Harrington instrumentation to correct the scoliosis; during this procedure the vascular supply to the spinal cord might be stretched. There were, however, no neurological abnormalities after operation in any of our

patients, all of whom had Harrington instrumentation. All of our patients have been seen regularly after leaving hospital and there have been no deteriorations in academic performances to suggest ischaemic damage to cerebral function. Fahmy, Mossad and Milad (1979) found that, in dogs, the effect of reduced blood pressure on the blood flow to the spinal cord and cerebrum was similar. Autoregulation of the cerebral blood flow in man occurs down to a mean arterial pressure of 40 millimetres of mercury (Conway 1978). It has been reported that sodium nitroprusside acts as a cerebral vasodilator and that in animals cerebral blood flow is well maintained at low levels of mean arterial pressure with this drug (Michenfelder 1980). Larsen *et al.* (1982), using sodium nitroprusside in anaesthesia during operations for cerebral aneurysm, found no change in cerebral blood flow down to a mean arterial pressure of 50 millimetres of mercury. A safe lower limit for mean arterial pressure in healthy patients under hypotensive anaesthesia has not

been universally agreed, but Cottrell (1982) suggests 50 millimetres of mercury and Khambatta *et al.* (1978) using a hypotensive technique similar to ours have reduced the mean arterial pressure to 42 millimetres of mercury without adverse effects. We feel that during operations for scoliosis the blood pressure must be accurately monitored and the mean arterial pressure kept above 50 millimetres of mercury.

In conclusion we can state that induced hypotension during posterior spinal fusion for idiopathic scoliosis is an effective technique for reducing blood loss during and after operation. Using this technique, a 55 per cent reduction in the blood requirement was achieved. Blood is an expensive and finite resource and any saving of donor blood is a major advantage. The method described is safe, providing the anaesthetist is experienced and the blood pressure is accurately monitored. No long-term or short-term impairment of cerebral or spinal function has been observed.

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LUQUE ROD INSTRUMENTATION IN THE TREATMENT OF ADOLESCENT IDIOPATHIC SCOLIOSIS

A COMPARATIVE STUDY WITH HARRINGTON INSTRUMENTATION

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Adolescent idiopathic scoliosis in 152 patients was treated by Luque L-rod instrumentation and early mobilisation without external support. This series was compared with a matched group of 156 patients treated by Harrington instrumentation and immobilised in an underarm jacket for nine months.

All the operations in both groups were performed by one surgeon and the patients were followed prospectively for more than two years. Correction of the scoliosis in the frontal plane was similar in both groups. However, the normal sagittal contour was better maintained with Luque rods, especially in the thoracolumbar and lumbar regions, and provided less loss of correction than with Harrington rods. Neither method significantly derotated the scoliosis. All the patients with Luque instrumentation developed a solid fusion despite breakage of the sublaminar wires at one or two levels in 4.9%. There were no major neurological complications with either type of instrumentation.

Luque L-rod instrumentation with segmental sublaminar wiring has been widely adopted in the treatment of neuromuscular scoliosis because it provides rigid fixation and allows early mobilisation without external support (Luque 1982a,b; Boachie-Adjei et al 1989; Broom, Banta and Renshaw 1989). However, its use in adolescent idiopathic scoliosis remains controversial, and there is concern regarding the surgical risks and long-term effects of passing wires into the spinal canals of patients who have no neurological abnormality (Wilber et al 1984; Thompson et al 1985). In contrast, Harrington instrumentation is a well-established method of treatment for idiopathic scoliosis, and when combined with a posterior fusion sets the standard against which any new method must be compared (Harrington 1962; Goldstein 1969; Harrington and Dickson 1973; Leider, Moe and Winter 1973; Erwin, Dickson and Harrington 1976; Livallo, Banta and Renshaw 1986). Any new technique must therefore provide better correction of the deformity, maintain spinal balance, produce a solid fusion and, most importantly, achieve these results with few complications.

The purpose of this paper is to compare the performance of Luque L-rod instrumentation, using segmental sublaminar wiring, with Harrington instrumentation in two matched groups of patients with adolescent idiopathic scoliosis undergoing posterior spinal fusion.

MATERIAL AND METHODS

Between 1975 and 1987, 308 patients with adolescent idiopathic scoliosis, all under the age of 19 years, were treated surgically at the Princess Margaret Rose Orthopaedic Hospital by means of posterior spinal fusion and instrumentation. These patients were studied prospectively and fell into two consecutive groups depending on the method of surgical instrumentation used to correct and stabilise the deformity. All the operations were performed by the author providing a uniform technical proficiency.

One group consisted of 152 patients treated between 1983 and 1987 by Luque L-rod instrumentation with segmental sublaminar wiring followed by early mobilisation without external support. The other group of 156 patients treated was between 1975 and 1983 by Harrington instrumentation followed by nine months in an underarm plaster jacket. The groups were well matched for age and sex and for the site and severity of the scoliosis.

All patients were followed for more than two years after the operation. The indications for surgical treatment were a patient over the age of 11 years with a scoliosis of

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more than 50° when first seen or a curve of more than 40° which was deteriorating.

Luque group. In this group there were 123 girls and 29 boys; their mean age at operation was 14 years seven months (range 11 years seven months to 18 years seven months). There was a single structural curve in 130 cases of which 105 were thoracic and 25 thoracolumbar. There were 22 patients with double structural scolioses; 20 were in the thoracic and lumbar regions, and two were double thoracic curves. The mean curvature, before operation, was 57° (range 40° to 115°). The mean follow-up time was three years two months (range two to six years).

All the operations were performed in the same way. The spine was exposed through a midline posterior incision and the muscles stripped subperiosteally to the tips of the transverse processes on both sides. The scoliosis was corrected by applying two pre-bent (4.76 mm diameter) stainless steel rods, one on either side of the spinous processes, throughout the length of the deformity. The two rods were bent intra-operatively to conform not only to the estimated degree of correction of the scoliosis, but also to create or maintain the normal thoracic kyphosis and lumbar lordosis, as well as to derotate the spine. The rods, which were L-shaped at one end to prevent migration up and down the spine, were fixed to each vertebra by means of 16-gauge (1.22 mm) stainless steel wires passed beneath the laminae and twisted on top of the rod. Double wires were used on both sides to fix the rods at their upper and lower ends. Passage of the wires required a midline laminotomy and excision of the ligamentum flavum at each vertebral level. The convexly curved rod was applied first and used to lever the spine straight by progressively wiring it to each of the vertebrae within the curve, starting from the upper neutral vertebra and proceeding to the lower neutral vertebra. Intra-operative spinal cord monitoring of somatosensory evoked potentials with an epidural electrode was used routinely from 1986.

A posterior spine fusion was performed throughout the instrumented area, but with some modifications compared to the Harrington group. The laminae were not decorticated because this could weaken the bone and cause the wires to cut through. The facet joints were excised bilaterally prior to the application of the spinal instrumentation, and once the spine was corrected they were packed with autogenous iliac bone grafts. More iliac bone, cut into matchsticks, was laid in both paraspinous gutters lateral to the Luque rods and overlying the excised facet joints and the decorticated transverse processes.

On the fourth day after surgery, patients were allowed to walk without any form of external support. They returned to school within two to three weeks and were allowed to swim after three months. They were told to avoid all rough sports for nine months.

Harrington group. In this group there were 123 girls and 33 boys. Their mean age at operation was 14 years six

months (range 11 years two months to 18 years ten months). There was a single structural curve in 128 cases of which 117 were thoracic and 11 were thoracolumbar. There were 28 patients with a double structural scoliosis of which 23 were in the thoracic and lumbar regions and five were double thoracic curves. The mean curvature before surgery was 60° (range 40° to 122°). The mean follow-up time was three years six months (range two years two months to seven years eight months).

The spine was exposed in the same manner as in the Luque group, but in these patients the scoliosis was corrected by applying a single Harrington distraction rod across the concavity between the neutral vertebrae at either end of the curve (Harrington 1962). In a double scoliosis the single rod spanned both curves. A posterior facet joint fusion was performed bilaterally in the same manner as in the Luque group, but in addition the laminae and transverse processes were deeply decorticated before the application of the iliac bone grafts (Moe 1958; McMaster 1980).

One week after surgery, a well moulded underarm plaster jacket was applied and the patient was allowed to walk. The jacket was worn for nine months, after which the patient was allowed unrestricted activities without external support.

Prophylactic antibiotics were given to both groups of patients commencing immediately before surgery, and continuing for 24 hours postoperatively.

RESULTS

The severities of the scoliosis, thoracic kyphosis and lumbar lordosis were measured, using the Cobb method, on anteroposterior and lateral spinal radiographs taken with the patient standing, before and after surgery, and then every six to nine months until the final assessment. The loss of correction was the difference between the angle measured on the immediate postoperative radiograph and that measured at the last examination.

Rotation of vertebrae was difficult to measure on the postoperative anteroposterior radiographs because the overlying rods obscured the concave margin of the vertebral body which is normally used as a reference point (Nash and Moe 1969; Pedriolle 1979). This problem was overcome by measuring the distance between the convex border of the apical vertebral body and the medial border of the pedicle on the same side, and comparing this with the width of the lower neutral vertebra at its mid-point. The lateral margins of the neutral vertebrae were not obscured by the rods and this ratio was used to estimate any change in the degree of rotation of the apical vertebra. Its use also overcame the problem of different magnifications of the serial radiographs.

For the purposes of analysis, both groups were divided into those with a single thoracic, single thoracolumbar, double (thoracic and lumbar) and double thoracic curves. Each curve type was subdivided into four, those

with curves from 40° to 49° from 50° to 59°, from 60° to 69° and those with curves of more than 70°. These subgroups were compared for differences between Luque and Harrington instrumentation regarding the angle of curve after correction, percentage correction and loss of correction.

This data, which is summarised in Tables I and II, was subjected to statistical analysis using standard *t*-tests and analysis of variance.

Single thoracic curves. Of the 222 single thoracic curves, 105 were treated by Luque L-rod instrumentation and 117 by Harrington instrumentation. A mean 10.5 vertebrae (range 9 to 13) per patient were instrumented in the

Luque group and 10.6 vertebrae (range 8 to 13) per patient in the Harrington group.

The pre-operative mean curve was 56° (range 40° to 107°) in those treated by Luque instrumentation and 61° (range 40° to 122°) in those who had Harrington instrumentation ($p = 0.03$). After surgery the mean curve in the Luque group was reduced to 25° (range 5° to 53°), a 56% mean correction (Figs 1a,b). In the Harrington group the mean curve after surgery was 28°, a 55% mean correction. There was no significant difference in the degree of correction obtained in the different sizes of curve nor between the two methods of instrumentation (Tables I and II).

Table I. Details of 152 patients with adolescent idiopathic scoliosis treated by Luque rod instrumentation

Type and curve severity*	Number	Mean age (yr mth)	Number of vertebrae fused	Mean curve*		Correction (per cent)	Mean loss of correction*
				Pre-operative	Postoperative		
Thoracic							
40 to 49	39	14 5	10.3	44	18	59	1.8
50 to 59	31	14 7	10.5	54	24	56	1.7
60 to 69	18	15 4	10.5	64	30	53	2.6
> 70	17	13 9	11.2	81	36	56	1.9
Thoracolumbar							
40 to 49	16	15 6	10.6	44	14	69	2.4
50 to 59	3	15 9	11	50	16	69	2.7
60 to 69	3	15	10.3	63	28	55	2.3
> 70	3	16	11.3	92	44	53	4.3
Double; thoracic and lumbar							
40 to 49	6	15	13.3	45:47	25:20	44:57	2:4.8
50 to 59	2	14 7	13	54:56	33:26	40:54	1:2
60 to 69	4	15 5	13	60:60	35:24	42:60	1.5:5.5
> 70	8	14 5	13.5	76:73	42:32	45:56	3:5
Double; thoracic	2	13 9	13	40:56	30:27	25:52	2:2

*measurement in degrees

Table II. Details of 156 patients with adolescent idiopathic scoliosis treated by Harrington instrumentation

Type and curve severity*	Number	Mean age (yr mth)	Number of vertebrae fused	Mean curve*		Correction (per cent)	Mean loss of correction*
				Pre-operative	Postoperative		
Thoracic							
40 to 49	36	14 1	10	43	19	57	4.5
50 to 59	24	14 5	10	54	23	58	5.4
60 to 69	22	15 2	10.5	64	30	54	5.3
> 70	35	14 10	10.5	83	39	53	5.5
Thoracolumbar							
40 to 49	5	15 3	10.5	43	16	61	9.5
50 to 59	3	15	11.5	55	18	64	10
60 to 69	1	18	13	61	20	67	10
> 70	2	15 7	9	79	36	55	13
Double; thoracic and lumbar							
40 to 49	4	14 3	11	43:42	21:16	51:62	8:9
50 to 59	8	13 6	12.75	54:46	26:18	52:61	8:8
60 to 69	3	14 5	12.3	60:63	32:31	47:51	5:9
> 70	8	15	13.1	74:75	43:37	42:51	9:10
Double; thoracic							
50 to 59	3	13 7	13	50:59	26:25	48:58	9:10
60 to 69	2	14 6	13	47:64	32:36	32:44	5:9

*measurement in degrees

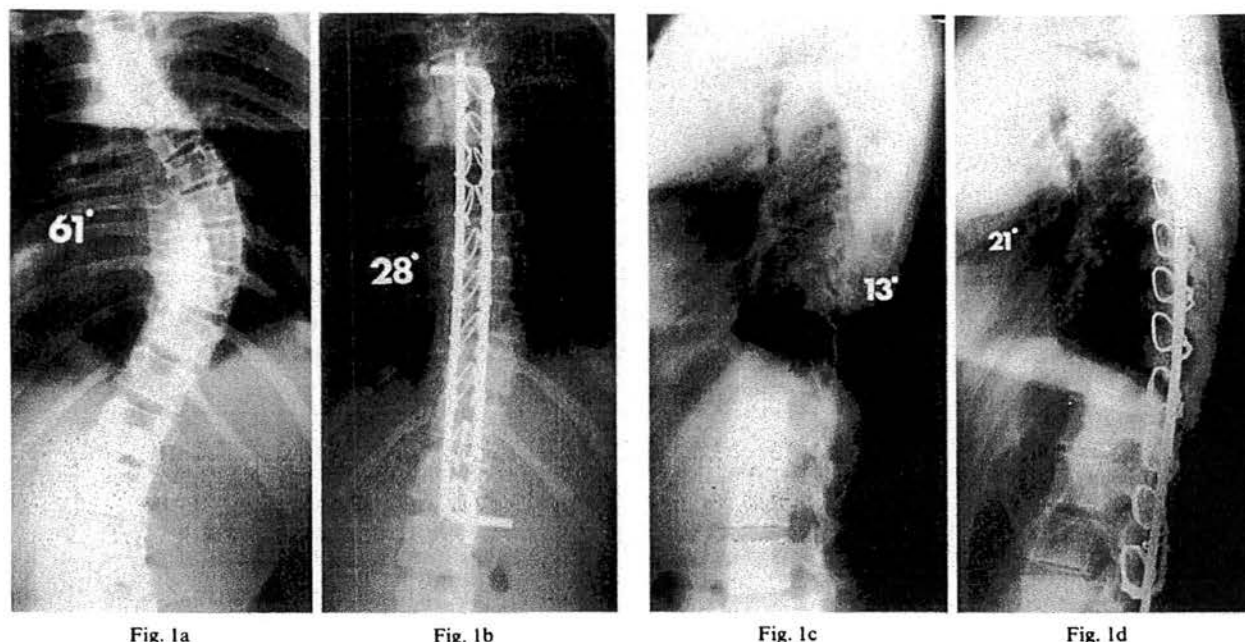


Figure 1a - A girl aged 14 years six months with a 61° adolescent idiopathic thoracic scoliosis; b) three years after Luque L-rod instrumentation and posterior fusion from T4 to L1, the curve remains well corrected at 28° and there is solid fusion; c and d) the pre-operative thoracic lordosis of 13° has been corrected to a 21° kyphosis.

The mean loss of correction in thoracic curves treated by Luque instrumentation was 1.9°, significantly less than the mean 5.1° lost in those treated by Harrington instrumentation ($p < 0.001$). In both treatment groups loss of correction was independent of the pre-operative size of the curve ($p = 0.74$, $p = 0.43$; Tables I and II).

The Luque rods were bent differentially in an attempt to derotate the vertebrae and also to maintain or restore the normal thoracic kyphosis. Despite this technique there was no significant change in the degree of vertebral rotation after surgery. However, the thoracic kyphosis which measured a mean 24° (range -9° to 58°) over the instrumented area before surgery was increased following Luque instrumentation by more than 5° in 28 patients (Figs 1c,d), remained unchanged in 56 and was decreased by more than 5° in 21 patients. If the kyphosis was less than 20° pre-operatively there was a tendency for it to increase (25 of the 35 patients) whereas in the 21 patients with a pre-operative kyphosis greater than 30°, 12 decreased and the remainder were unchanged.

Harrington instrumentation had no effect on derotating the spine and also caused a flattening of the thoracic kyphosis in all the patients. Before surgery the mean thoracic kyphosis over the instrumented area was 26° (range -7° to 48°) and this was significantly reduced to a mean 12° (range 10° to 28°) after surgery.

Thoracolumbar curves. Of the 36 thoracolumbar curves, 25 were treated by Luque L-rods and 11 by Harrington rods. A mean of 10.7 vertebrae (range 8 to 13) per patient were instrumented in the Luque group and 10.4 vertebrae (range 9 to 13) per patient in the Harrington group. The lower ends of the Luque rods extended to L3 in four

patients, L4 in 15 and L5 in six; the Harrington rods extended to L4 in all 11 patients.

The pre-operative mean curve was 53° (range 40° to 115°) in those treated by Luque instrumentation and was 53° (range 40° to 83°) in those with Harrington instrumentation. After surgery, the mean curve in the Luque group was reduced to 19° (range 3° to 60°), a 65% mean correction (Figs 2a,b). The degree of correction was similar with the Harrington instrumentation; the mean curve was reduced to 20°, a 62% correction ($p = 0.46$). However, with the Luque, but not with the Harrington instrumentation, there was significantly greater correction of curves of less than 60° compared to those of greater angle (Tables I and II).

The mean loss of correction for thoracolumbar curves treated by Luque instrumentation was 2.7°; this ranged from 2.4° for pre-operative curves of less than 50° to 4.3° for curves of over 70°. In the Harrington group the mean loss of correction ranged from 9.5°, for curves of 40° to 49°, to 13° for curves of more than 70°. The mean loss (10.1°) was significantly greater than in the group treated with Luque rods ($p = 0.0003$).

The pre-bent Luque rods were successful in maintaining or restoring the lumbar lordosis in all 25 patients (Figs 2c,d). Before surgery the mean lumbar lordosis over the instrumented area was 18° (range 15° kyphosis to 36° lordosis) and following Luque instrumentation this was improved to a mean 21° (range 2° to 48°). However, rotation of the spine was improved in only 15 patients and remained unchanged in ten.

Harrington instrumentation had no effect on rotation and caused flattening of the lumbar lordosis in all the

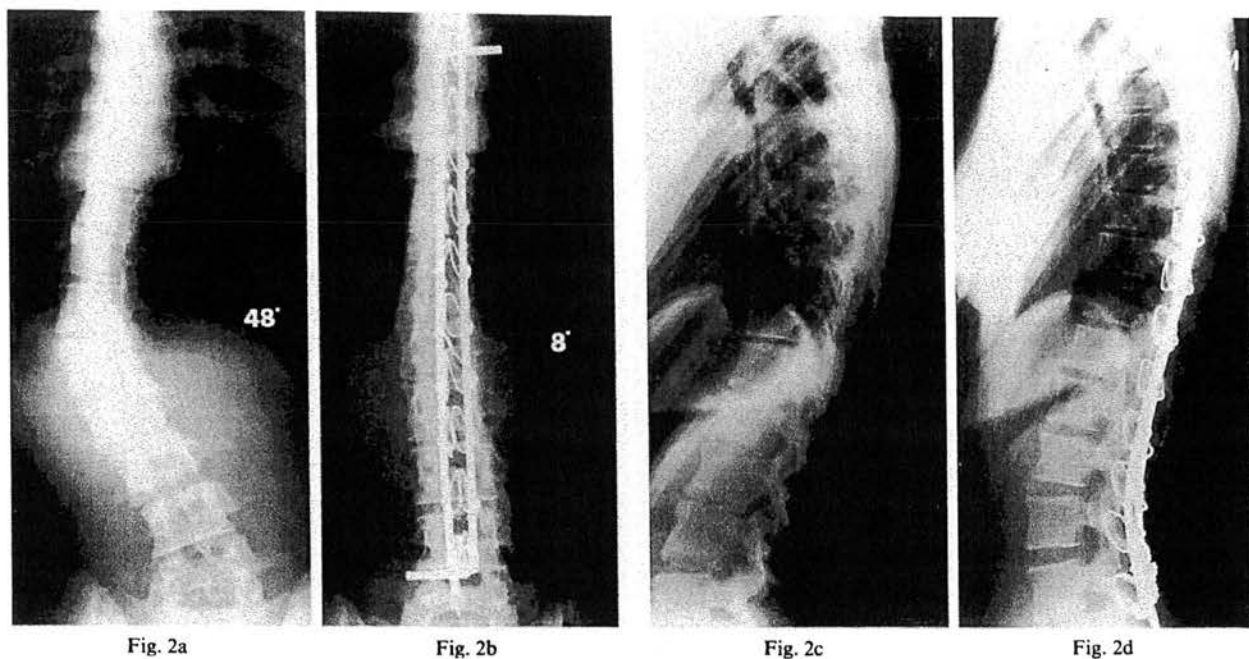


Figure 2a – A girl aged 15 years one month with a 48° adolescent idiopathic thoracolumbar scoliosis; b) two years and ten months after Luque L-rod instrumentation and posterior fusion from T7 to L4, the curve remains well corrected at 8° and there is solid fusion; c and d) The pre-operative lumbar lordosis has been well maintained.

patients. Before surgery the mean lumbar lordosis over the instrumented area was 20° (range 7° kyphosis to 35° lordosis) and this was significantly reduced to 8° (range 8° kyphosis to 14° lordosis) after surgery.

Double curves, thoracic and lumbar. Of the 43 double structural curves in the thoracic and lumbar regions, 20 were treated with Luque rods and 23 with Harrington rods. A mean 13.3 vertebrae (range 12 to 15) per patient were instrumented in the Luque group and 12.6 vertebrae (range 11 to 15) per patient in the Harrington group ($p = 0.025$). The lower ends of the Luque rods extended to L4 in 19 patients and to L5 in one; the Harrington rods extended to L3 in two patients, to L4 in 15 and to L5 in six patients (Tables I and II).

The pre-operative mean thoracic and lumbar curves were 61° and 62° respectively in the Luque group and 60° and 58° in the Harrington group.

The effect of Luque instrumentation was different for each of the two curves (Figs 3a,b). The thoracic curves corrected in the same way as did the single thoracic scolioses; there was a 43% correction in the frontal plane, no change in rotation and in only ten curves (50%) was there improvement by more than 5° in the sagittal plane. The lumbar curves corrected in the same manner as did the single thoracolumbar scolioses; there was a mean 56% correction in the frontal plane and the lumbar lordosis was maintained in all the patients, but only 40% had an improvement in rotation.

With Harrington instrumentation, the mean correction of the thoracic and lumbar curves in the frontal plane was 47% and 56% respectively, not significantly

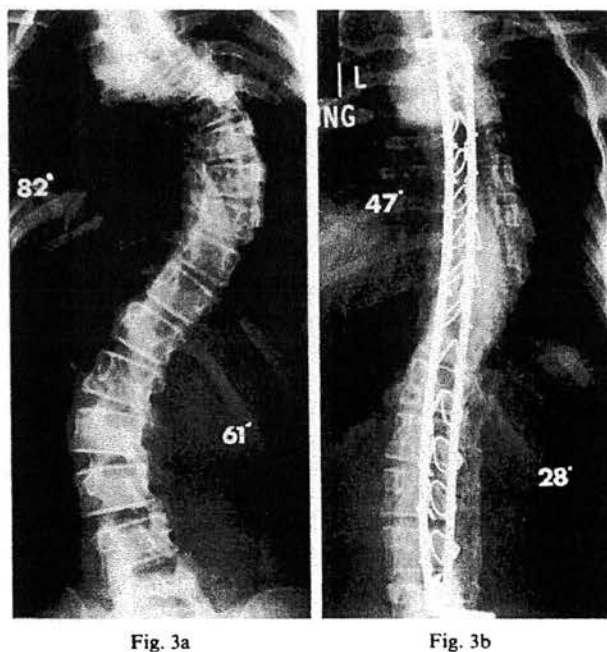


Figure 3a – A girl aged 15 years with a double structural adolescent idiopathic scoliosis; b) two years six months after surgery good correction has been maintained and there is a solid fusion.

different from the correction obtained by Luque instrumentation. However, the main disadvantage of Harrington rods was significant flattening both of the thoracic kyphosis and the lumbar lordosis, so that the spine became one straight column in the sagittal plane over

the instrumented area. Vertebral rotation remained unchanged.

The mean loss of correction for the thoracic and lumbar curves treated by Luque rods was 2.2° and 4.7° respectively. In the Harrington group the mean loss of correction, 8° and 9.3° was significantly greater.

Double thoracic curves. Of the seven patients with double structural thoracic curves, two were treated with Luque rods and five with Harrington rods (Tables I and II). There were too few for significant comparison.

Operative time and blood loss. The mean duration of the operations on patients treated with Luque rods was 195 minutes (range 165 to 225); and in the Harrington group, 180 minutes (range 120 to 240). The mean total blood loss, during and after the operation, in the Luque group was 1490 ml (range 695 to 2945) and in the Harrington group, 1200 ml (range 500 to 2458).

Duration of hospital stay. The mean time spent in hospital for the Luque treated patients was 13 days (range 11 to 21), significantly less than the 19 days (range 11 to 45) required for the Harrington group.

COMPLICATIONS

Neurological. There were no major neurological complications as a result of either method of instrumentation. However, four patients treated with Harrington rods (2.5%) and one patient treated with Luque rods (0.6%) complained of mild paraesthesia affecting the soles of the feet; the patients with Harrington instrumentation had curves which had all been corrected by more than 60% whereas the one patient with Luque instrumentation had only 45% correction. There were no objective neurological abnormalities and the paraesthesia resolved completely within 24 to 48 hours in all five patients. A further two patients treated with Luque rods (1.3%) complained of mild paraesthesia extending in a radicular manner around one side of the trunk; this also recovered completely within a few months.

A mild brachial plexus palsy occurred in two patients; one with Luque and one with Harrington instrumentation. Both patients were noted to have paraesthesia and slight weakness of one hand which was thought to be due to stretching of the lower roots of the brachial plexus caused by hyperabduction of the arm during the operation. Both recovered normal power and sensation within a few months.

Paraesthesia in the distribution of the lateral cutaneous nerve of the thigh occurred in two patients with Harrington instrumentation. This was thought to be due to pressure on the nerve by the plaster jacket and recovered once the plaster was trimmed.

Wound healing. One patient who had Luque instrumentation had a major wound infection. This presented several weeks after the operation as a discharging sinus in the upper part of the wound and did not heal with antibiotics. Five months later the sinus was explored and

found to extend to the fusion mass which was noted to be solid with no pseudarthroses. The sinus was completely excised and the Luque rods and sublaminar wires were removed. The wound then healed and when last seen, 40 months later, there was only a 2° loss of correction. There were no major infections in the patients with Harrington instrumentation, but two developed superficial infections of the iliac crest wounds which responded rapidly to local care and antibiotics.

Minor wound dehiscence, which required resuture, occurred in two patients in each treatment group.

Haematomas occurred in 13 patients with Luque instrumentation (8%), two in spinal wounds and 11 in iliac crest wounds. They were thought to be due to the very early mobilisation of these patients and did not occur so frequently in patients who were kept in bed for four days after operation. Eight patients with Harrington instrumentation (5%) developed haematomas, three in spinal wounds and five in iliac crest wounds.

Failure of instrumentation. None of the Luque rods fractured, but the sublaminar wires broke at one or two levels in seven patients (4.6%). There was a total of ten broken wires out of the 3330 sublaminar wires employed (0.3%). The wires broke at the lower two instrumented levels in seven patients and also at the uppermost level in two. However, none of these patients had any related symptoms or neurological complications; all developed a solid fusion, and in only one was there a significant loss of correction. This patient had a 42° thoracolumbar curve which was corrected to 7°. Within the first three months the wires broke at the lower level where they secured the junction of the L-rod and the straight rod, allowing the rods to rotate so that the lordotic curve which had been pre-bent into the rods became a scoliotic curve. The deformity relapsed to 22°. This patient, who has been followed for 29 months, has had no further loss of correction. Surgery was never necessary to remove the broken wires in any of the patients.

One patient with Harrington instrumentation was found to have a broken rod at radiographic review three years seven months after operation. She had lost 10° of correction, but there was a solid fusion and no treatment was thought necessary. Six years later there was no further loss of correction. Two patients with double thoracic scolioses treated with Harrington rods dislocated the upper hooks at the T1 level within a week of operation. Both required a second operation to re-insert the hooks, which were then stabilised with methyl-methacrylate cement.

Pseudarthrosis. There were no pseudarthroses in the patients with Luque instrumentation. One patient with Harrington rods (0.6%) developed a pseudarthrosis. This was a 13-year-old girl with a thoracolumbar scoliosis which had been corrected from 45° to 8°. Six months after operation there was a 9° loss of correction, and a pseudarthrosis was diagnosed radiographically at the thoracolumbar junction. The pseudarthrosis was repaired

and a plaster jacket applied for five months, after which the spine became solidly fused. The patient has been followed for three years, and there has been no further loss of correction.

DISCUSSION

This study and others have shown that Harrington rods are an effective means of achieving correction of idiopathic scoliosis in the frontal plane (Harrington and Dickson 1973; Leider, Moe and Winter 1973; Erwin, Dickson and Harrington 1976; Tolo and Gillespie 1981; Lovullo et al 1986), but that the method does not derotate the spine and that it produces flattening of the normal sagittal contour (Casey et al 1987; Luk et al 1987). Cochran, Irstam and Nachemsom (1983) found that if the instrumentation extended to the lower lumbar region, the reduction in lordosis could increase the stress on the unfused distal segments and predispose to backache and degenerative changes in later life. On occasion there may be an excessive flattening of the lumbosacral spine resulting in the 'flat back' syndrome in which the patient is tilted forwards and only able to stand erect by flexing the knees (LaGrone et al 1988). The ideal form of instrumentation should not only correct the lateral curve, but should maintain or restore the normal thoracic kyphosis and lumbar lordosis as well as derotating the spine.

In this study Luque L-rod instrumentation and Harrington instrumentation were found to be equally effective in correcting the frontal plane deformity regardless of the site or degree of the curve. However, Luque instrumentation was significantly more effective in maintaining correction. A further advantage was that the contoured Luque rods were much more effective in preserving the normal sagittal profile of the spine, especially in the thoracolumbar and lumbar regions. The major defect of the Luque system was that it had no effect on derotating the vertebrae in the thoracic region, and had only a moderate effect in the thoracolumbar and lumbar regions.

The relative failure of Luque instrumentation to simultaneously derotate as well as correct the spine in the frontal and sagittal planes may be explained by the three-dimensional nature of the original deformity. In idiopathic thoracic scoliosis there is a short segment lordosis at the apex of the curve and the axis of rotation lies posterior to these vertebrae (Dickson et al 1984). It is therefore only possible to simultaneously derotate and restore the normal kyphosis by pulling the concave side of the vertebrae directly backwards towards this axis. Luque instrumentation is ineffective because the convex rod, which is pre-bent to restore the thoracic kyphosis, is applied first and used to lever the spine straight (in the frontal plane) by wiring it to each of the vertebrae within the curve. Although this may maintain the thoracic kyphosis, it cannot derotate the spine. Once the convex

rod is in position the spine is fixed, and it is not possible to use the concave rod to pull the concave side of the spine backwards, and so derotate the vertebrae. However, in the thoracolumbar and lumbar regions, the Luque rods are more effective in correcting the deformity in all three planes. The convex rod which is used to straighten the spine in the frontal plane is pre-bent anteriorly to maintain or create the normal lumbar lordosis, and as this rod is levered into position it pushes the convexity of the spine forward, and so effects a degree of vertebral rotation as well as preserving the lumbar lordosis.

The risk of neurological complications following Harrington instrumentation in adolescent idiopathic scoliosis is approximately 0.5% (MacEwan, Bunnell and Sriram 1975). With Luque rods there is an added risk due to the passage of the wires into the spinal canal, and there is a significant learning curve. The incidence of neurological complications following Luque instrumentation in adolescent idiopathic scoliosis has been reported to be as high as 17%, with major cord injury occurring in 4% (Herring and Wenger 1982; Moore and Eilert 1983; Wilber et al 1984; Thompson et al 1985). An American Scoliosis Research Society survey (Morbidity and Mortality Committee Report 1987) found a four times greater risk of spinal cord injury following sublamina wiring than with standard Harrington instrumentation. Bunch and Chapman (1985) found that patients about to undergo spinal surgery were most concerned by the risk of neurological complications, followed by the fear of having to have a second operation. They were less concerned about the need to wear a brace or cast, and about the amount of correction obtained. In these circumstances, it is debatable whether the advantages of Luque rods outweigh their possible risks. However, in this series, all the sublamina wires were passed by one experienced surgeon and there were no serious neurological complications.

It was necessary to remove the Luque rods and wires in one patient six months after surgery because of a major wound infection. Fortunately, this too was performed without neurological complications, but removing multiple sublamina wires embedded in bone is riskier than removing Harrington rods and hooks and this is a major disadvantage of the Luque technique.

An advantage of the Luque system over Harrington rods is that it provides fixation at each level so that loss of fixation at one level, due to bone failure or wire breakage, does not necessarily compromise the whole system. No further operations were necessary in the seven patients in whom sublamina wires broke, whereas two patients with Harrington instrumentation required further surgery to relocate dislocated hooks.

The long-term success of the surgery of spinal deformity depends not on the instrumentation, but on the achievement of a solid spinal fusion. If a pseudarthrosis develops, any form of implant will ultimately fail and correction will be lost. It has been shown that a solid

fusion is more likely to occur following meticulous bilateral interfacet and intertransverse fusion, followed by decortication of the laminae and the onlay of large amounts of autogenous iliac bone grafts (Moe 1958; Goldstein 1969; Erwin et al 1976; McMaster 1980). With Luque instrumentation the area of bone available for fusion is much smaller than with Harrington instrumentation, and it is not safe to decorticate the laminae because it weakens the fixation. However, none of the patients in this series with Luque instrumentation was thought to have developed a pseudarthrosis, although the radiographic diagnosis is not easy when the fusion mass is obscured by an overlying metal implant. Furthermore, the very rigid fixation provided by Luque rods may prevent loss of correction, which is often the only indication of a pseudarthrosis. Though all the patients were followed for more than two years, it is still possible for a pseudarthrosis to manifest itself later by the development of a fractured rod.

In conclusion, Luque rod instrumentation with segmental sublaminar wiring is a more difficult and potentially dangerous procedure. However, with care and experience, good results can be achieved with few complications, though the long-term effect of wires within the spinal canal remains unknown. The main advantages of Luque rods are that they maintain the normal sagittal contour, especially in the thoracolumbar and lumbar regions, and prevent loss of correction better than Harrington rods. The greater rigidity provided by segmental sublaminar wiring allows the patient to be mobilised within a few days of operation without external support. The main disadvantage of the method is that it has very little effect on derotating the spine, especially in the thoracic region.

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The Effect of Luque Segmental Sublaminar Instrumentation on the Rib Hump in Idiopathic Scoliosis

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The change in back shape after Luque segmental sublaminar instrumentation was assessed in the frontal, sagittal, and transverse planes in 61 patients with adolescent idiopathic scoliosis using the Integrated Shape Investigation System (ISIS) and standard radiographic techniques. Luque instrumentation was found to be an effective method of correcting thoracic and thoracolumbar curves in the frontal plane with a 59% and 63% respective reduction in the size of the preoperative Cobb angle. Despite the frontal plane correction, however, the ISIS scan showed that of the 40 single thoracic curves, the rib hump was reduced in only 6 patients, was unchanged in 27 patients, and was worsened slightly in 7 patients. By contrast, thoracolumbar and lumbar curves were corrected in all three planes with a significant cosmetic improvement. [Key words: adolescent idiopathic scoliosis, Luque segmental sublaminar instrumentation, rib hump correction, Integrated Shape Investigation System, ISIS]

ADOLESCENT IDIOPATHIC SCOLIOSIS is a complex spinal deformity which occurs in the frontal, sagittal, and transverse planes. The spine not only curves to the side but also rotates about its long axis, producing a unilateral prominence of the trunk. The rib cage is elevated posteriorly on the convex side of the curve and depressed on the concave side; it is often the rib hump rather than the lateral curve which is the major cosmetic deformity.

These patients are usually girls who are concerned about their appearance, and the ideal surgical treatment should attempt to improve their back shape by correcting the deformity in all three planes. In the past the results of surgery have usually been assessed using anteroposterior and lateral spinal radiographs which only estimate correction in the frontal and sagittal planes but give no indication of a change in the transverse plane with an increase or decrease in the unilateral rib prominence. Attempts to measure this prominence have been made using a spinal pantograph¹⁴ and contour devices such as the formulator^{1,10} and the flexicurve.⁹ Unfortunately these methods are not very accurate for small changes and are subject to operator error when used serially. The Integrated Shape Investigation System (ISIS, Oxford Metrics Ltd., Oxford, UK) was developed more recently and accurately measures the surface shape of the patient's back in three dimensions using a computerized stereophotogrammetric technique.^{11,13} This is an easily repeatable method which is subject to minimal operator error and provides an objective means of monitoring even small changes in back shape. We assessed the change in back

shape after Luque instrumentation in patients with adolescent idiopathic scoliosis using ISIS and standard radiographic methods.

MATERIALS AND METHODS

Since April 1985, ISIS has been used routinely in the Edinburgh Scoliosis Unit to assess prospectively the back shape of all patients with adolescent idiopathic scoliosis before and after corrective spinal surgery. The system consists of a light source, closed-circuit video camera, computer, and plotter. The patient stands in a frame which ensures accurate positioning. A narrow horizontal beam of light scans down the patient's back in synchrony with the video system, and this information allows an accurate three-dimensional map of the back to be created by the computer in the length of time it takes to scan the back (less than 2 seconds). The stance of the patient is corrected automatically by the computer in the transverse and frontal planes with respect to markers applied to the posterior superior iliac spines and the spinous process of the seventh cervical vertebra. Markers are also placed at regular intervals along the line of the spinous processes.

The map of the back is displayed by a series of ten horizontal "bacon slices," which are horizontal sections at equal distances from the cervicothoracic junction to the sacrum. A full analysis is available in the form of a printout a few minutes after the patient is scanned.

During the period April 1985 to July 1987, 61 patients with adolescent idiopathic scoliosis underwent a posterior spinal fusion with correction of their deformity using the Luque method of segmental sublaminar spinal instrumentation.⁵ There were 53 girls and 8 boys (mean age at surgery, 14 years 8 months; range, 10 years 8 months to 21 years 7 months). Of these patients, 40 had thoracic curves, 14 had thoracolumbar curves, and 7 had double structural curves affecting the thoracic and lumbar regions.

All patients had their spinal deformity assessed before surgery and between 3 and 9 months after surgery using radiographs, the ISIS technique, and clinical photography. Standardized anteroposterior and lateral spinal radiographs of the whole spine taken with the patient standing were used.

The surgery was done by one surgeon (MJM) using a standard technique. Two Luque 3/16-in. diameter rods were prebent to correct not only the lateral deviation of the spine but also, where appropriate, to create or maintain a physiologic thoracic kyphosis or lumbar lordosis. In addition the rods were bent differentially to try and correct vertebral rotation. The convex rod was always applied first and was used as the main correcting device. The posterior spinal fusion and Luque rods extended from the upper to the lower neutral vertebrae, and all of the vertebrae within the curve were secured to the rods by bilateral segmental sublaminar wiring. The mean number of vertebrae instrumented and fused in the thoracic and thoracolumbar curves was 10 (range, 9-13), and in double curves affecting the thoracic and lumbar regions, it was 13 (range, 12-15). After surgery the patients were allowed to walk on postoperative day 4 and sent home without external support between days 10 and 14. They returned to normal school after a further 2-3 weeks and avoided sports and contact activities until the spine was solidly fused at 9 months.

ISIS Assessment. An ISIS scan of the patient's back shape was done the day before surgery and 3 months later. The ISIS technique is based on the surface shape of the back; therefore much of the information given by scans done immediately after surgery was not as reliable as at other times. This was because of the hematoma and soft tissue swelling that occurred immediately postoperatively and quickly subsided.

The line joining the spinous markers is used by ISIS to estimate the volumetric asymmetry of the surface of the back preoperatively. Postoperatively the line of the scar is normally used for the placement of markers because the spinous processes are removed at surgery. This does not correspond to the true line of the spine and calculated values of volumetric asymmetry using these marker placements are subject to unacceptable errors. For this reason the size of the rib hump in the thoracic region or the muscle prominence in the lumbar region was assessed by the angle of trunk rotation, ATR, measured directly from the horizontal sections on the printout from the scans using A3 paper. The angle between the line marking the frontal plane and a line across the posterior aspect of the back was measured for each section and the maximum quoted as the ATR. The ISIS corrects the scan so that the frontal plane passes parallel to the posterior superior iliac spines and the spinous process of the seventh cervical vertebra. This measurement is similar to the angle of inclination measured by Burwell et al.¹

A change in the ATR of 4° or more after surgery was regarded as significant, and these patients were graded better, worse, or unchanged with regard to their transverse plane deformity (Table 1).

Thoracic translocation is also a deforming factor and is the lateral displacement of the patient's chest to the convex side of the curve relative to the pelvis. This accentuates the prominence of the contralateral hip and flattens the waist and hip on the same side. The severity of thoracic translocation was measured on the ISIS scan by the following technique: 1) a line was drawn between the markers on the posterior superior iliac spines, 2) a second line was drawn vertically up from the midpoint of this line, 3) the horizontal distances from the vertical line to the outline of the back were measured at the apex of the thoracic curve, and 4) the difference in these two distances (quoted as a percentage of the width of the outline at the same level) gives the thoracic translocation. A difference of more than 5% was regarded as abnormal.

Spinal height was measured directly from the ISIS scan with a line running from the marker on the spinous process of the seventh cervical vertebra to the midpoint between the markers on the posterior superior iliac spines. The calculation of the spinal height from the ISIS scan required calculation of the midpoint of the line joining the markers and then calculating the distance from this point to the marker on the spinous process of the seventh cervical vertebra. It was believed that measurement of this distance on an A3 plot was as accurate as the calculated distance.

Radiographic Assessment. The severity of the scoliosis, thoracic kyphosis, and lumbar lordosis were measured on the anteroposterior and lateral spinal radiographs using the Cobb method. Decompensation of the upper trunk in relation to the pelvis was measured on the standing anteroposterior spinal radiographs by dropping a perpendicular with respect to the top of the radiograph through the spinous process of C7 and projecting the line downward to the sacrum. Normally this should pass through the sacrum. Rotation of the apical vertebra could not be measured directly on the postoperative anteroposterior spinal radiographs because the overlying Luque rods obscured the concave margin

of the vertebral body. It was not possible, therefore, to measure the vertebral width which is normally used to estimate vertebral rotation.^{7,8}

A new technique was developed in which the minimum width of the lower neutral vertebra was used, because these are not obscured by the Luque rods. The distance was measured from the medial border of the pedicle of the apical vertebra on the convex side of the curve to the vertebral border on the same side, and the ratio of this distance to the width of the neutral vertebra was recorded. This measurement can be used to measure the change in rotation of the apical vertebra on serial radiographs with the same accuracy as other techniques.

RESULTS

Deformity in the frontal plane was assessed radiographically by measuring the Cobb angle, the thoracic decompensation, and the thoracic translocation from the ISIS scan. A change in the spinal height also reflected a change in the frontal plane. Transverse plane deformity was assessed by the ATR measured from the ISIS scan and radiographically by the vertebral rotation. Sagittal plane deformity was measured from the radiographs using the Cobb method.

Single Thoracic Curves

Frontal Plane. There were 40 patients with a single thoracic scoliosis whose mean Cobb angle before surgery was 58° (range, 40–84°). After Luque instrumentation, this was reduced to a mean of 23° (range, 8–47°) a mean improvement of 59% ($P < 0.001$ by paired Student *t* test).

There was a varying degree of thoracic translocation in all of the 40 patients with a thoracic scoliosis (mean, 19%; range, 4–44%) as measured on the ISIS scan. Five of these patients had a thoracic translocation of less than 5%, taken to be within the normal range, and they were unchanged after surgery. Thirty-five patients had a thoracic translocation of greater than 5% (range, 6–44%), and of these, 23 improved to within the normal range, 9 improved but not to normal (range, 6–12%), and 3 were unchanged (range, 6–8%). No patient became worse. The spinal height, as measured on the ISIS scan, improved by a mean of 7.2% (range, 0–17%) after surgery.

Transverse Plane. The size of the rib hump as measured by the ATR on the ISIS scan was reduced in only 6 patients (Figures 1A–F), was unchanged in 27 patients, and was worsened slightly in 7 patients (Figures 2A–F). Before surgery the mean ATR in the 40 patients was 8° (range, 2–17°). In the six patients whose rib hump was reduced by surgery, the mean ATR decreased from 11° to 6°. In the seven patients whose rib hump was made worse, the ATR increased from a mean of 6° to 10°.

The Luque rods were bent in an attempt to derotate the vertebrae in the curve and also maintain or restore the normal thoracic kyphosis. The mean rotation of the apical vertebra in the curves before surgery was 22° (range, 7–38°), and after surgery, there was no change in any of the patients.

Sagittal Plane. The mean thoracic kyphosis before surgery was 20° (range, 5–58°), and after surgery, this was improved by more than 5° (range, 6–18°) in 14 patients, was unchanged in 16 patients, and decreased by greater than 5° in 10 patients. If the preoperative kyphosis was less than 10°, there was a tendency for it to increase in size (9 of 12 patients). If the kyphosis was greater than 25°, it either was unchanged or decreased in size after surgery (Table 2). In addition there was no correlation between a change in the thoracic kyphosis and a change in the size of the rib hump.

Thoracolumbar Scoliosis

Frontal Plane. There were 14 patients with a single thoracolumbar scoliosis whose mean Cobb angle before surgery was 49° (range,

Table 1. Effect of Luque Instrumentation on the Rib Hump

Type of Curve	No. of Patients	Better	Unchanged	Worse
Thoracic	40	6	27	7
Thoracolumbar	14	13	1	0
Double				
Upper curve	7	0	6	1
Lower curve	7	4	1	2

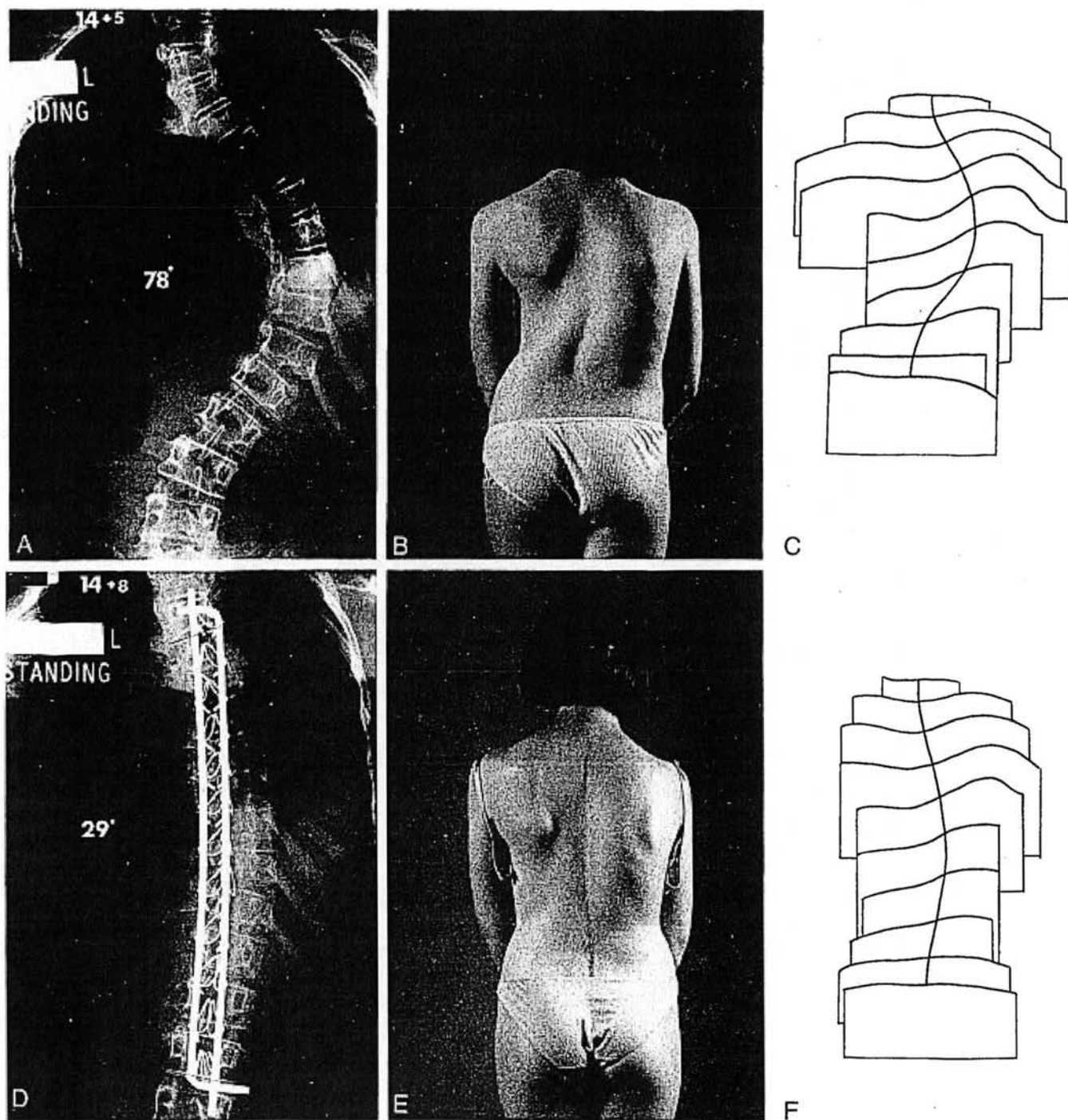


Fig 1. A–F, A 14-year, 5-month-old girl with a thoracic scoliosis measuring 78°. Postoperatively this corrected to 29°, with a reduction in the size of the rib hump as measured by the ISIS scan.

40–88°). After Luque instrumentation, this was reduced to 17° (range, 6–43°), a mean improvement of 63% ($P < 0.001$).

Decompensation (or listing) of the trunk, as measured on the standing anteroposterior radiographs occurred in all 14 patients (range, 3–14 cm), and after surgery, this improved to within less than 1 cm of normal in all but one patient. Thoracic translocation was present before surgery in all patients (mean, 22%; range, 10–40%). After surgery this

decreased to a mean 6% (range, 0–16%); 9 patients were normal, 3 improved, and 2 were unchanged. The spinal height (measured on the ISIS scan) improved by a mean 7% (range, 0–15%) after surgery.

Transverse Plane. The rib hump was reduced in 13 of the 14 patients, mainly due to an elevation of the rib depression on the concave side of the curve (Figures 3A–F). Before surgery the mean ATR in these

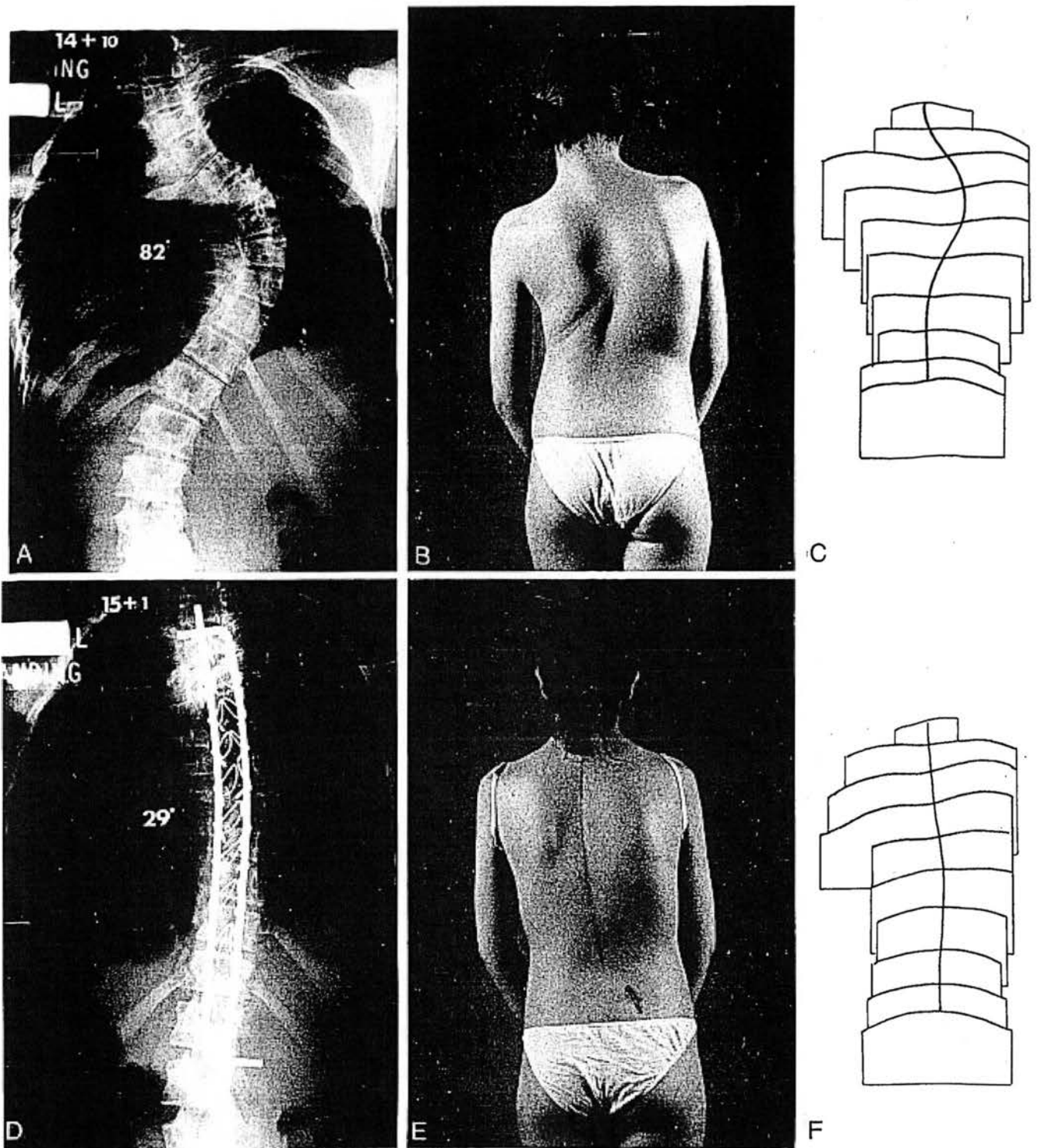


Fig 2. A-F, A 14-year, 10-month-old girl with a thoracic scoliosis measuring 82°. Despite a correction to 29° and an improved cosmetic appearance, the rib hump increased in size.

13 patients was 14° (range, 8–21°), and after Luque instrumentation, this was reduced to 5° (range, 0–12°). Only one patient was unchanged with a rib hump of 11°.

The Luque rods were bent in an attempt to derotate the vertebrae in the curve and also maintain the normal lumbar lordosis. Despite the reduction in the size of the rib hump after surgery, the apical vertebral

rotation decreased in only four patients and was unchanged in ten patients.

Sagittal Plane. In this plane the instrumentation was more successful in maintaining the lumbar lordosis (Figure 4). The mean lumbar lordosis as measured over the instrumented levels from T12 to L4 before

Table 2. Effect of Luque Instrumentation on Kyphosis in Thoracic Curves

Preoperative Kyphosis (°)	No. of Patients	Decreased	Unchanged	Increased
10	12	0	3	9
10-20	8	0	4	4
20-25	6	1	4	1
25	14	9	5	0

surgery was a mean 27° (range, 12–46°). After Luque instrumentation, the mean lumbar lordosis was minimally changed at 29° (range, 12–44°), and no patient changed for better or worse by more than 5°.

Double Scoliosis

Frontal Plane. Seven patients had double structural curves affecting the thoracic and lumbar regions. Before surgery the mean thoracic scoliosis was 59° (range, 42–77°), and the mean lumbar scoliosis was 54° (range, 40–67°). This was reduced to a mean 33° (range, 19–51°)

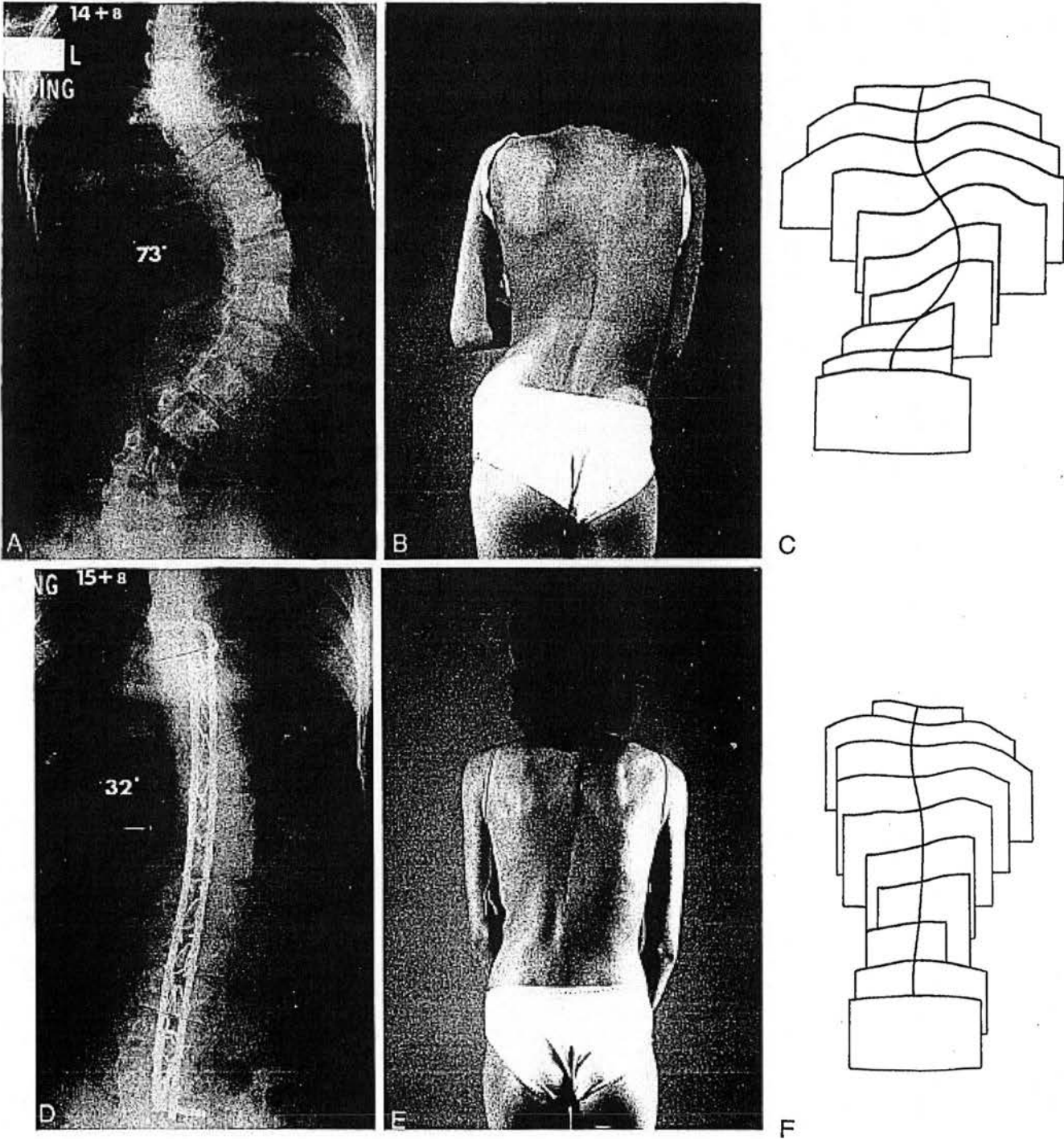


Fig 3. A–F, A 14-year, 8-month-old girl with a thoracic lumbar scoliosis measuring 73° preoperatively. Postoperatively the curve measured 82°, and there was a marked improvement in appearances and reduction in the size of the rib hump.

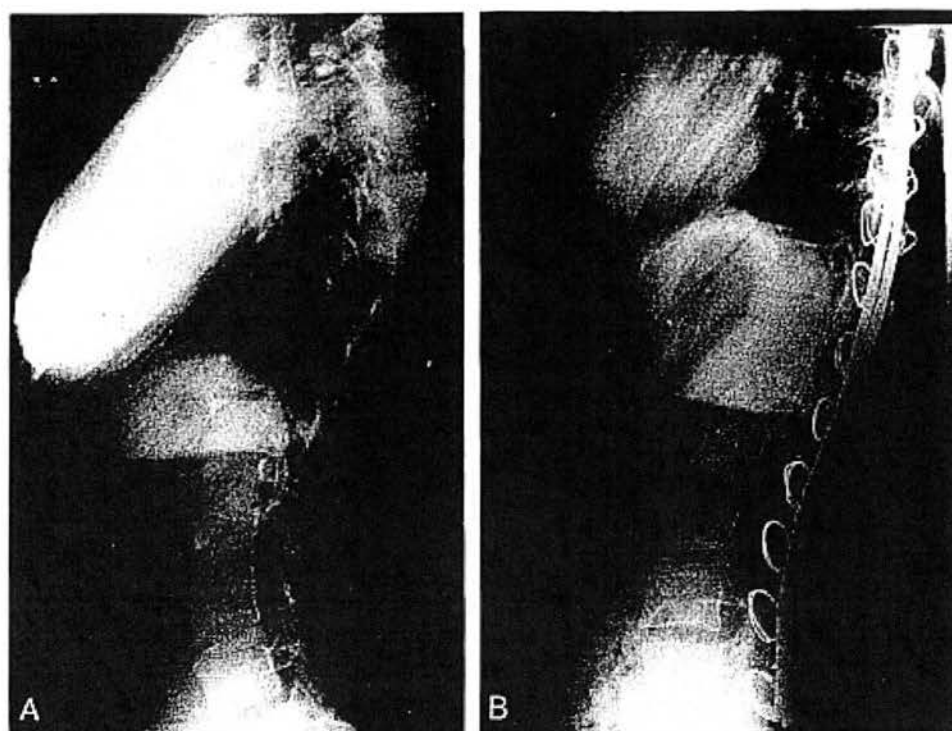


Fig 4. Pre- and postoperative lateral radiographs showing maintenance of the lumbar lordosis after Luque segmental spinal instrumentation.

for the thoracic curves and a mean 23° (range, $14\text{--}40^\circ$) for the lumbar curves, a mean improvement of 44% and 57%, respectively. The thoracic translocation was normal in all except two patients before surgery, and these normalized postoperatively. Spinal height increased by a mean 6% (range, 0–10%).

Transverse Plane. The size of the rib hump associated with the thoracic curve was a mean 7° (range, $1\text{--}23^\circ$) before surgery. After surgery this was unchanged in six patients and increased in one patient from 9° to 13° . In no patient was the rib hump decreased by the surgery. The lateral muscle prominence in association with the lumbar curve was a mean 8° (range, $4\text{--}18^\circ$) before surgery. After surgery this decreased in four patients to a mean 3° (range, 0– 8°), was unchanged in one patient, and increased from a mean of 4° to 9° in two patients.

Sagittal Plane. The thoracic kyphosis (before surgery a mean of 21° with a range of $8\text{--}26^\circ$) increased by greater than 5° (range, $6\text{--}23^\circ$) in four patients and was unchanged in the remaining three patients. The lumbar lordosis (before surgery a mean of 31° with a range of $18\text{--}55^\circ$) was not changed by more than 5° for better or worse in any of the patients.

DISCUSSION

This study using the ISIS technique has shown that the effect of Luque segmental sublamina spinal instrumentation on the back shape of patients with idiopathic scoliosis depends on the site of the scoliosis. Thoracic curves were all significantly corrected in the frontal plane after Luque instrumentation, but only a minority improved in the sagittal plane and very few in the transverse plane. In the frontal plane, the Cobb angle was corrected by 59%, the spine lengthened by 7%, and the thoracic translocation improved to normal in 85% of the patients. In the sagittal plane, the tendency to thoracic hypokyphosis usually found in idiopathic scoliosis was improved in 35% of the patients. Despite these improvements in the frontal and sagittal planes, the attempt to derotate the vertebral column and trunk in the transverse plane by differentially

bending the Luque rods was usually unsuccessful: Only 15% of patients had a reduction in the size of their rib hump, and the degree of vertebral rotation was unchanged in all patients. On reviewing the clinical photographs, however, the rib hump appeared to be improved in all of the patients, and this was thought to be due to the increase in spinal height which spread the deformity over a larger area and made it seem less obvious.

In contrast, thoracolumbar curves could be corrected in all three planes with a significant cosmetic improvement. In the frontal plane, the Cobb angle was corrected by 63%, the spine lengthened by 7%, the thoracic translocation improved to normal in 93% of the patients. The tendency to list corrected to normal in all but one patient. In the transverse plane, the size of the rib hump was significantly reduced in 13 of the 14 patients, due, in part, to an elevation of the depression on the side of the concavity of the curve and, in part, to a reduction in the height of the rib hump on the convexity. Despite this improvement in the size of the hump, the degree of vertebral rotation was reduced in only 4 of the 14 patients. In the sagittal plane, the physiologic lumbar lordosis was maintained in all patients, and this prevented the development of the "flat back" syndrome which can occur after the insertion of a straight Harrington rod to correct a scoliosis in the region.^{4,6} This is a very disabling condition in which there is a flattening of the normal lumbar lordosis resulting in the patient being tilted forward and not being able to compensate by hyperextending the hips.

In double structural curves affecting the thoracic and lumbar regions the effect of Luque instrumentation was different for each type of curve. The thoracic curves tended to correct in the same manner as for a single thoracolumbar scoliosis; they improved in the frontal and transverse planes, and the physiologic lumbar lordosis was maintained in the sagittal plane.²

This study has shown that Luque segmental sublamina spinal instrumentation is an effective method of correcting thoracolumbar adolescent idiopathic scoliosis in all three planes, but thoracic curves improved significantly only in the frontal plane. These results for thoracic curves are very similar to those found by Weatherley et al,¹²

who used a formulator device to study the rib hump in patients with adolescent idiopathic scoliosis before and after Harrington instrumentation. They found that Harrington instrumentation did not have any corrective effect on the thoracic rib hump. After 1 year 36% of the patients had a rib deformity greater than preoperatively, and after 2 years, this had increased to 64%. In this study using Luque instrumentation, 13% of the patients had a rib hump slightly greater than before surgery, and it is possible that these numbers could increase with further follow-up.

In idiopathic scoliosis there appears to be no direct relation between correction of the thoracic lateral curvature using either Harrington or Luque instrumentation and reduction in the size of the rib hump or a reversal of the degree of vertebral rotation. This failure to achieve correction in all three planes simultaneously may be explained partially by the three-dimensional nature of the scoliosis.^{2,3} Thoracic curves are associated with a lordotic segment at the apex of the curve, and the axis of vertebral rotation lies posterior to this region. The spine therefore cannot be derotated by pushing or pulling the vertebrae directly toward the midline by segmentally wiring the vertebrae within the curve to rods which lie anterior to the axis of spinal rotation. Theoretically the vertebral column can only be derotated by segmentally pulling the concave aspect directly posteriorly toward the axis of spinal rotation. Biomechanically Luque instrumentation is ineffective in correcting thoracic vertebral rotation because the convex rod is applied first and used to lever the spine straight by progressively wiring it to each vertebra in the curve, starting from the upper neutral vertebra and proceeding to the lower neutral vertebra. It is not possible for this rod to derotate the vertebrae and maintain or create a physiologic thoracic kyphosis. Once the convex rod is in position, the spine is fixed, and it is no longer possible to use the concave rod to pull the concave aspect of the vertebrae backward and so derotate the spine and trunk. In contrast, Luque instrumentation is biomechanically much more effective in improving the back shape in thoracolumbar scoliosis because the convex rod is used not only to straighten the scoliosis but is also prebent anteriorly to maintain or create a physiologic lumbar lordosis. When the convex rod is applied to the spine, it pushes the apex of the convexity of the curve forward and reduces the rib hump although the degree of vertebral rotation does not appear to be significantly affected. In thoracic curves the reverse applies. The convex rod is prebent backward to restore the thoracic kyphosis, and this is much more likely to increase the size of the rib hump.

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The Effect of Luque-Rod Instrumentation on the Sagittal Contour of the Lumbosacral Spine in Adolescent Idiopathic Scoliosis and the Preservation of a Physiologic Lumbar Lordosis

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The changes in the sagittal alignment of the lumbar spine were investigated in 28 patients with adolescent idiopathic scoliosis undergoing long posterior spinal fusion to L4 or L5 with contoured Luque rods and segmental sublaminar wiring. The lumbar lordosis over the instrumented levels was preserved, and there was no compensatory hyperlordosis of vertebral segments distal to the fusion. In situations where long posterior spinal fusions are indicated, instrumentation with contoured Luque rods and segmental sublaminar wiring can preserve the normal sagittal alignment of the lumbosacral spine. [Key words: long posterior spinal fusion, adolescent idiopathic scoliosis, Luque-rod instrumentation, sagittal alignment]

PATIENTS WITH ADOLESCENT idiopathic scoliosis may present with thoracolumbar or double major curves extending into the lower lumbar region necessitating treatment by means of a long posterior fusion extending distally to L4 or L5. Surgical correction of these curves using Harrington distraction instrumentation will produce a flattening of the sagittal contour of the instrumented lumbar spine and the possible development of a compensatory hyperlordosis below the instrumented levels at the lumbosacral junction.^{3,7} Long-term studies of these patients have revealed a high incidence of low-back pain,^{1,4} and it has been suggested that the increased stress on the unfused distal lumbosacral segment may predispose the patient to back pain and the development of degenerative changes. On occasion, there may be an excessive flattening of the lumbar spine resulting in a loss of sagittal balance and a "flat back syndrome"⁹ in which there is a fixed, forward inclination of the trunk and an inability to stand erect when the knees are fully extended.⁶ It is therefore important that any form of spinal instrumentation extending into the lower lumbar region maintain the normal physiologic lumbar lordosis.

The purpose of this study is to assess the effect of contoured Luque-rod (L-rod) instrumentation on the sagittal contour of the lumbosacral spine in patients with adolescent idiopathic scoliosis who had undergone long posterior spinal fusion to L4 or L5.

MATERIALS AND METHODS

From 1983 to 1987, 144 patients with adolescent idiopathic scoliosis were treated at the Princess Margaret Rose Orthopaedic Hospital, Edinburgh, by a posterior spinal fusion and correction by means of double L-rod instrumentation. In 36 patients, the fusion extended to L4 or L5, and preoperative and postoperative radiographs were available in 28. There were 4 boys and 24 girls whose mean age at surgery was 16.5 years (range, 11–19 years). Fifteen patients had a single thoracolumbar curve, and 13 patients had double major curves affecting the thoracic and lumbar regions.

Double L-rods ($\frac{3}{16}$ -in diameter) were applied in parallel and bilaterally fixed the spine by segmental sublaminar wiring (16-gauge) at every level from the upper to the lower neutral vertebra. These rods were contoured not only to correct the scoliotic deformity in the coronal plane, but also to preserve the normal lumbar lordosis in the sagittal plane throughout the instrumented area. The instrumentation extended to L4 in 24 patients and to L5 in 4 patients.

Measurements were made on the preoperative and postoperative standing lateral spinal radiographs of the total lumbar lordosis, the lordosis over the instrumented lumbar levels, the lordosis distal to the instrumented levels, and the sacrohorizontal angle (Figures 1–2). The total lumbar lordosis (Figure 1[A]) is the angle between the superior end plate of T12 and the top of the sacrum. The lordosis in the instrumented area (Figure 1[B]) is the angle between the superior end plate of T12 and the superior end plate of L4 or L5, depending on the extent of the instrumentation. Similarly, the lordosis below the instrumented levels (Figure 1[C]) is the angle between the superior end plate of the most distally fused vertebra (L4 or L5) and the top of the sacrum. The sacrohorizontal angle (Figure 1[D]) is measured by the method of Wiltse and Winter.¹⁶ Statistical differences in the preoperative and postoperative sagittal plane measurements were determined with the Student *t* test. Linear regression analysis was used to examine the correlation between changes in the measured parameters.

Measurements were also made on the preoperative and postoperative standing anteroposterior spinal radiographs of the thoracolumbar or lumbar scoliosis by the use of the Cobb method.

RESULTS

The 15 patients with a single thoracolumbar curve had a mean preoperative scoliosis of 49° (range, 35–73°), and this was reduced to 15° (range, 3–31°) after L-rod instrumentation—a mean improvement of 69% (Table 1).

The 13 patients with a double major curve had a mean preoperative thoracic scoliosis of 60° (range, 42–87°) and a mean lumbar scoliosis of 57° (range, 43–100°). After L-rod instrumentation, these curves were reduced to 32° (range, 25–46°) and 23° (range, 14–46°), respectively—a mean improvement of 47% and 60%, respectively (Table 1).

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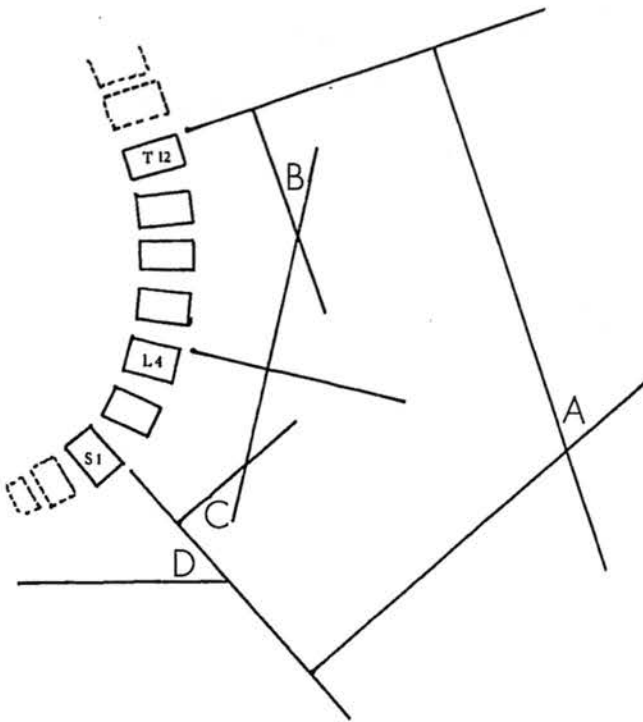


Fig 1. Line diagram to illustrate the method of measurement of lordosis of the lumbosacral spine in the sagittal plane. **A:** Total lumbar lordosis between the superior end plate of T12 and the superior end plate of S1. **B:** The lumbar lordosis over the instrumented level between the superior end plate of T12 and the superior end plate of L4. **C:** The lordosis of the lumbar spine below the instrumented levels between the superior end plate of L4 and the superior end plate of S1. **D:** The sacrohorizontal angle measured by the method of Wiltse and Winter.¹⁶

Table 1. Patients

	Scoliosis (°)		Improvement (%)
	Preoperative	Postoperative	
Single thoraco lumbar curve (n = 15)	49 (35-73)*	15 (3-31)	69
Double thoraco lumbar curves (n = 13)			
Upper	60 (42-87)	32 (25-46)	47
Lower	57 (43-100)	23 (14-46)	60

* Figures in parentheses indicate the ranges of values.

The mean total lumbar lordosis in the 28 patients before surgery was 49° (range, 17-77°); after L-rod instrumentation this was increased slightly to a mean of 54° (range, 22-106°), which was not statistically significant ($.5 > P > .1$) (Table 2).

The mean lumbar lordosis over the instrumented area before surgery was 18° (range, 15° of kyphosis to 36° of lordosis); after L-rod instrumentation, this was also slightly increased to a mean of 21° (range, 2-48° of lordosis), which was not significant. After surgery, no patients had a kyphosis over the fused segment. In 9 of the 28 patients, however, the lordosis over the instrumented area was increased by 10° or more. In this subgroup, the mean preoperative lumbar lordosis over the instrumented area was only 9° (range, 15° of kyphosis to 26° of lordosis) and included three patients in whom the lumbar spine was kyphotic. This degree of lumbar "lordosis" was significantly less than the mean of the remaining 21 patients (mean, 22°; $P > .02$). In these nine patients, the lordosis over the instrumented levels was increased after surgery, to a mean of 30° (range, 15-48° of lordosis).

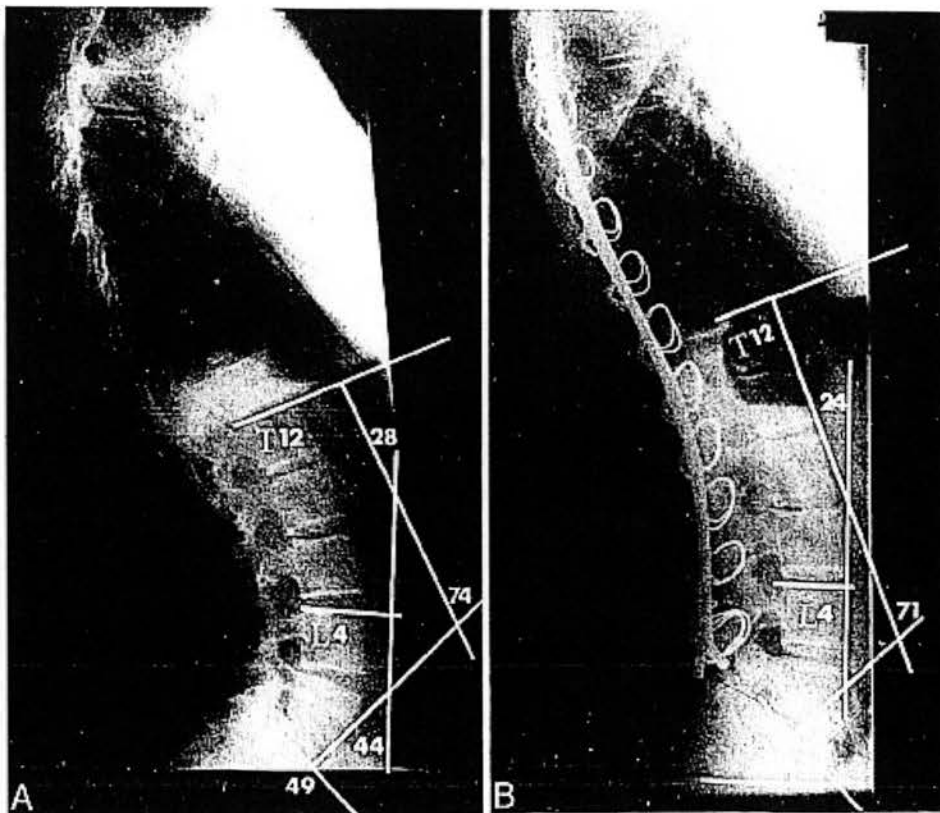


Fig 2. Preoperative (**A**) and post-operative (**B**) lateral radiographs of a patient undergoing long posterior spinal fusion and instrumentation with L-rods and sublaminar wiring for adolescent idiopathic scoliosis to L4. The preoperative sagittal configuration of the spine has been well preserved. The thoracolumbar scoliosis was corrected from 47° before surgery to 10° after surgery, as measured by the Cobb method on the antero-posterior radiograph.

Table 2. Mean Measurements* in the Sagittal Plane of the Lumbar Spine for All Patients

	Preoperative	Postoperative
Total lumbar lordosis	49.5 ± 15.2 (17-77)†	54.5 ± 15.4 (22-106)
Lordosis over fused levels	17.9 ± 13.5 (+15-36)‡	21.1 ± 12.0 (2-48)
Lordosis distal to fusion	31.4 ± 10.5 (14-47)	33.3 ± 13.1 (7-74)
Sacrohorizontal angle	39.2 ± 9.5 (15-55)	39.0 ± 12.1 (19-80)

*In degrees ± SD.

†Figures in parentheses indicate range of values.

‡A positive (+) notation indicates a kyphosis; all unmarked values indicate a lordosis.

Conversely, six patients had a loss of lumbar lordosis over the instrumented levels of 10° or more after surgery. In this subgroup, the mean lordosis before surgery was significantly greater ($P > .001$) at 31° (range, 25–36°) compared to the lumbar lordosis over the instrumented levels in the remaining 22 patients (mean, 14°). After surgery, the lumbar lordosis in this small subgroup of six patients was reduced to a mean of 14° (range, 4–25°).

The mean lumbar lordosis distal to the instrumented area before surgery was 31° (range, 14–47°); after surgery, this was minimally increased to a mean of 33° (range, 7–74°), which was not statistically significant. In 8 of the 28 patients, however, lordosis distal to the instrumented area was increased by 10° or more. In only three of these eight patients was there a loss of lordosis over the instrumented level of 10° or more. There was a poor correlation between the change in the lumbar lordosis over the instrumented area and the lordosis distal to it in the lumbosacral region ($r = 0.335$); $.1 > P > .05$.

There was no significant alteration in the mean value of the sacrohorizontal angle after surgery (39.2° and 39.0° before and after surgery, respectively).

Patients undergoing instrumentation to L4 or L5 were separated into two groups; the values of the measured parameters are shown in Table 3. The small number of patients ($n = 4$) undergoing fusion to L5 precludes accurate statistical comparison, but there seemed to be a greater increase in the lordosis of the fused segment and no increase in the lordosis at the lumbosacral junction.

DISCUSSION

It is now widely accepted that a spinal fusion in patients with adolescent idiopathic scoliosis should, if possible, not extend into the lower lumbar region.² In thoracolumbar and lumbar curves, stabilization with Zielke anterior spinal instrumentation preserves a greater number of mobile lower lumbar segments than stabilization with posterior spinal instrumentation.⁸ There are situations, however, in which it is not possible to avoid a long posterior fusion with instrumentation extending to L4 or L5. In these circumstances, Harrington instrumentation using either straight or contoured square-ended rods will result in a loss of lumbar lordosis over the instrumented levels,³ and this becomes more pronounced the more distal the fusion.^{1,4} Although the total lordosis may fall within a normal range, the instrumented upper lumbar spine is often flattened or even kyphotic,⁷ with a compensatory hyperlordosis of the unfused lower lumbar spine and extension of the sacrum.^{1,4} This may increase the stress on the remaining mobile segments and predispose the patient to the development of backache and early degenerative changes.¹⁴

Long-term studies have shown that the lower the level of Harrington

Table 3. Mean Measurements* in the Sagittal Plane of the Lumbar Spine Showing Those Instrumented to L4 (N = 24) and L5 (N = 4)

	Level of Instrumentation	
	L4	L5
Total lumbar lordosis		
Preoperative	50.0 ± 16.2 (17-77)†	46.3 ± 7.8 (35-52)
Postoperative	54.5 ± 16.2 (22-106)	54.0 ± 10.5 (45-69)
Lordosis over fused levels		
Preoperative	16.8 ± 13.8 (+15-36)‡	24.0 ± 11.1 (8-33)
Postoperative	18.8 ± 10.1 (2-35)	34.8 ± 14.4 (15-48)
Lordosis distal to fusion		
Preoperative	33.3 ± 9.3 (17-47)	22.3 ± 5.7 (14-27)
Postoperative	35.4 ± 11.8 (17-74)	19.3 ± 10.5 (7-29)
Sacrohorizontal angle		
Preoperative	38.8 ± 10.2 (15-55)	41.4 ± 2.7 (38-44)
Postoperative	38.4 ± 12.6 (19-80)	42.3 ± 9.4 (30-53)

*In degrees ± SD.

†Figures in parentheses indicate the range of values.

‡A positive (+) notation indicates a kyphosis; all unmarked values indicate a lordosis.

instrumentation, the higher the incidence of back pain.^{1,4} In contrast, Moskowitz et al¹⁰ reported no increase in the incidence of low-back pain 20 years after *in situ* fusions for adolescent idiopathic scoliosis in which the lumbar lordosis was well preserved. Wasylenko et al¹⁵ have also shown that the maintenance of a normal physiologic lumbar lordosis after surgery for spinal deformity is also important for energy consumption, ambulation, and cosmesis. Therefore, it is important that any form of spinal instrumentation extending into the lower lumbar region should not only correct the deformity in the frontal and axial planes, but also maintain a normal physiologic lumbar lordosis over the instrumented levels and a normal lumbosacral angle. The normal physiologic lordosis varies widely.^{12,13} In a group of 114 children with scoliosis, the mean lumbar lordosis was 48° (range, 40–55°),¹² which is similar to the patients presented here. The sacrohorizontal angle in the review of Stagnara et al¹³ was 41° (range, 19–65°), which is also similar to the value measured in our patients (39°, Table 2).

Any system using distraction instrumentation to correct a scoliosis will tend to flatten the spine in the sagittal plane. In the L-rod system, however, the rods are contoured to preserve the lumbar lordosis and are levered into position without significantly distracting the spine. This study is in agreement with others^{5,11} and has shown that such a system extending to L4 or L5 is capable of preserving and even restoring a normal lumbosacral sagittal alignment and does not produce a secondary hyperlordosis in the unfused segments distal to the instrumented levels or alter the sacral inclination.

The importance of the preservation of a physiologic lumbar lordosis with respect to the onset of degenerative changes and symptoms of low-back pain remains the subject of a long-term study.

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Segmental vessel ligation in patients undergoing surgery for anterior spinal deformity

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Segmental vessel ligation during anterior spinal surgery has been associated with paraplegia. However, the incidence and risk factors for this devastating complication are debated.

We reviewed 346 consecutive paediatric and adolescent patients ranging in age from three to 18 years who underwent surgery for anterior spinal deformity through a thoracic or thoracoabdominal approach, during which 2651 segmental vessels were ligated. There were 173 patients with idiopathic scoliosis, 80 with congenital scoliosis or kyphosis, 43 with neuromuscular and 31 with syndromic scoliosis, 12 with a scoliosis associated with intraspinal abnormalities, and seven with a kyphosis.

There was only one neurological complication, which occurred in a patient with a 127° congenital thoracic scoliosis due to a unilateral unsegmented bar with contralateral hemivertebrae at the same level associated with a thoracic diastematomyelia and tethered cord. This patient was operated upon early in the series, when intra-operative spinal cord monitoring was not available.

Intra-operative spinal cord monitoring with the use of somatosensory evoked potentials alone or with motor evoked potentials was performed in 331 patients. This showed no evidence of signal change after ligation of the segmental vessels.

In our experience, unilateral segmental vessel ligation carries no risk of neurological damage to the spinal cord unless performed in patients with complex congenital spinal deformities occurring primarily in the thoracic spine and associated with intraspinal anomalies at the same level, where the vascular supply to the cord may be abnormal.

The spinal cord is supplied by two different arterial systems: three longitudinal arterial trunks lying within the spinal canal, and the segmental arteries, arising from the aorta.¹ The longitudinal arterial trunks extend along the cord beneath the meninges from the medulla oblongata to the conus medullaris. They are the anterior median longitudinal spinal artery (anterior spinal artery) and the posterolateral spinal arteries. The arteriolar vessels that arise from these arteries encircle the spinal cord, forming the vasocorona, a fine pial plexus that provides limited anastomoses between the anterior and posterolateral spinal arteries.

Bilateral segmental vessels arise from the aorta and inferior vena cava and pass around both sides of each vertebra before entering the spinal canal through the intervertebral foramina.

The arteries divide before entering the foramina and provide the nutrient vessels to the vertebral bodies, as well as a rich anastomotic network on the surfaces of the vertebral

bodies, transverse processes and pedicles. These anastomoses are between ipsilateral and contralateral arteries. After entering the intervertebral foramen and penetrating the dura, the posterior branch of each segmental artery divides into dural arteries, which supply the spinal dura and root sleeve, and radicular arteries, supplying the anterior and posterior nerve roots. A second anastomotic network is formed in the extradural space within the spinal canal, with the greatest concentration in the cervical and lumbar regions. These anastomotic networks are believed to preserve spinal cord circulation by providing an alternative route for arterial flow during conditions of stress.

Also, the posterior branch of the segmental artery is the origin of the medullary feeders, which reinforce the longitudinal arterial channels in a sporadic fashion and at various levels. These medullary feeders are supplied mainly by the vertebral arteries in the cervical spine and by aortic segmental arteries in the thoracic

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Table I. Study population and type of surgery performed

Diagnosis	Number of patients	ASF* with instrumentation	ASF without instrumentation	A/PSF† with posterior instrumentation	A/PSF with A/P‡ instrumentation	A/PSF without instrumentation
Idiopathic scoliosis	173	64	6	95	4	4
Congenital spinal deformity	80	4	10	27	1	38
Neuromuscular scoliosis	43	1	2	4	31	5
Miscellaneous/syndromic scoliosis	31	3	4	11	2	11
Scoliosis associated with intraspinal abnormalities (neuropathic scoliosis)	12	2	0	7	0	3
Kyphosis (Scheuermann's/post-laminectomy)	7	0	0	6	0	1

* ASF, anterior spinal fusion

† A/PSF, anterior/posterior spinal fusion

‡ A/P, anterior/posterior

and lumbar regions. The medullary arteries enter the dura adjacent to the nerve root ganglion, after which they ascend and anastomose with an anterior or posterolateral spinal artery to supply the spinal parenchyma (medulla). The anterior spinal artery does not receive an afferent supply at each segmental level.

The medullary feeders, commonly labelled 'radicular arteries' reinforce the longitudinal arterial channels at various levels.¹ The largest medullary feeder of the lumbar cord is the arteria radicularis anterior magna (great radicular artery, or artery of Adamkiewicz), which in 80% of cases originates from a left segmental artery between T7 and L4, with a predilection for the T9 to T11 levels. Biglioli et al² reported the artery of Adamkiewicz to enter the spinal canal from the left side between T9 and L5 in 63% of patients, below T12 in 70%, and between L1 and L3 in 65%. These and other investigators have suggested that there may be more than one arteria radicularis anterior magna in 11% of patients.²⁻⁴

In 1974, Domisse¹ defined the area between T4 and T9 as the 'critical vascular zone of the spinal cord', recognising that at these levels the spinal canal was narrowest and the blood supply least profuse. He concluded that this was the zone in which interference with the spinal circulation was most likely to result in paralysis.

Paraplegia may occur following impairment of the segmental vessels. The incidence of paraplegia was 8% in a study of 138 patients undergoing resection of a thoracic or thoracolumbar aneurysm necessitating bilateral ligation of the segmental vessels without spinal cord monitoring.² This was reduced to 2% in a subsequent group of 95 patients using spinal cord monitoring, in whom the segmental vessels were first sequentially clamped and only ligated if there was no change in the somatosensory evoked potentials within eight to ten minutes of occlusion.³

Patients with severe or rigid spinal deformities often require anterior spinal surgery to release, decompress or fuse the spine, with or without internal fixation. This usually requires ligation of the segmental vessels over the length of the deformity. A vascular insult following disruption

of the blood supply to the cord is recognised as a possible factor leading to permanent neurological compromise in these patients.

The purpose of this study was to record the prevalence of neurological complications in a large number of paediatric and adolescent patients with spinal deformities undergoing anterior surgery performed through a thoracic or thoracoabdominal approach using the same technique of segmental vessel ligation throughout, and to assess any neurophysiological changes occurring as a consequence of ligating these vessels.

Patients and Methods

We reviewed the medical records and spinal radiographs of 346 consecutive paediatric and adolescent patients with a scoliosis or kyphosis who underwent anterior surgery through a thoracic or thoracoabdominal approach (Table I). Patients with myelomeningocele or spinal cord injury with pre-operative neurological deficits were excluded. The mean age at surgery was 12.3 years (3 to 18). The senior author (MJM) operated on 319 patients with the remaining 27 anterior or anteroposterior procedures performed by the first author (AIT) using the same technique of anterior approach and ligation of the segmental vessels.

There were 173 patients (50%) with an idiopathic scoliosis, 80 (23%) with a congenital scoliosis or kyphosis, 43 (12.5%) had a neuromuscular scoliosis, 31 (9%) had a syndromic scoliosis, 12 (3.5%) had a scoliosis associated with intraspinal abnormalities but without neurological deficits and seven (2%) had kyphosis. There were no cases of revision surgery. The deformity was thoracolumbar or lumbar in 240 patients (69%) and thoracic in 106 (31%).

All the patients with a congenital spinal deformity had a pre-operative myelogram or MR scan of the whole spine to assess the presence of intraspinal anomalies. There were 29 patients with a congenital scoliosis or a scoliosis associated with a Chiari malformation, diastematomyelia or other intraspinal anomaly. They were defined as having a 'spinal cord at risk' during surgery because of spinal dysraphism.

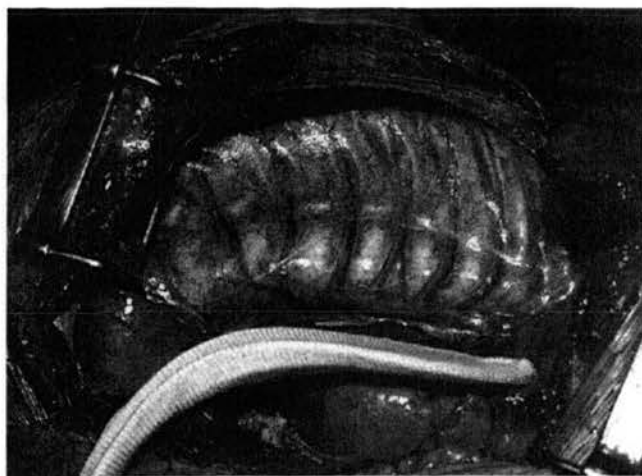


Fig. 1a



Fig. 1b



Fig. 1c

Intra-operative photographs which show our technique of a) and b) unilateral ligation of the segmental vessels with silk sutures at the area where they cross the mid-portion of the vertebral bodies, avoiding proximity with the intervertebral foramina. This is followed by c) a complete discectomy and annulectomy back to the posterior longitudinal ligament, as well as resection of the rib heads across the levels of the deformity.

A total of 250 patients (72.3%) underwent a combined anterior and posterior spinal fusion, with posterior instrumentation in 150 patients, anterior and posterior instrumentation in 38, and no instrumentation in the remaining 62. The other 96 patients (27.7%) had only an anterior spinal fusion, with instrumentation in 74 cases. A total of 2651 segmental vessels were ligated; a mean of 7.7 vessels per operation (6 to 9).

Early in the series, 15 patients underwent correction of scoliosis without intra-operative neurophysiological monitoring. In these patients spinal cord function was assessed after correction of the deformity using a wake-up test. Between 1985 and 1999, 168 patients were operated on using cortical or epidural somatosensory evoked potentials. Between 2000 and 2007, 163 patients underwent their surgery with the addition of motor evoked potential, to the

somatosensory (cortical and spinal) evoked potential recording. This provided a more complete neurophysiological evaluation of neural function during surgery, with the aim of detecting impending spinal cord injury while it remained potentially reversible.

A significant neurophysiological change, indicating potential or actual spinal cord damage, was defined as a reduction in the amplitude of the somatosensory evoked potentials by > 50% and/or altered latency of > 10% compared with the baseline, as well as intra-operative loss of the motor evoked potentials.⁵

Operative technique. The anterior procedure was performed in the lateral decubitus position, using a thoracic or thoracoabdominal retroperitoneal approach. Hypotensive anaesthesia was used in all cases, the mean blood pressure being maintained intra-operatively at 60 mmHg. After

exposure of the lateral aspect of the vertebral bodies on the convexity of the scoliosis, the segmental vessels were separately released and ligated with silk sutures where they crossed the mid portion of the vertebral body, avoiding proximity to the neuroforamen, where important collateral vessels may exist. The segmental vessels were never ligated on both sides at the same level. In none of our patients was soft clamping of the vessels before ligation performed.

Ligation of the segmental vessels was followed by excision of the rib heads and a complete annulotomy and discectomy back to the posterior longitudinal ligament, in order to allow angular and rotational mobility of the spinal segments (Fig. 1). The application of morcellised autologous rib graft enhanced anterior fusion across the excised discs.

Results

Only one patient had neurological deterioration after anterior spinal surgery. This was a 14-year-old girl with a left-sided congenital thoracic scoliosis measuring 127° due to a unilateral unsegmented bar extending from T8 to T12 with three contralateral hemivertebrae at the same level (T9 to T11). She also had a diastematomyelia at the T11 to T12 level, with tethering of the spinal cord but normal pre-operative neurology. She underwent neurosurgical excision of the bony spur and a de-tethering procedure without neurological complications. This was followed six months later by a two-stage anterior and posterior scoliosis correction. The first stage was a convex vertebral body resection and an osteotomy of the unilateral unsegmented bar performed through an anterior left thoracic approach to the spine on the convexity of the scoliosis. The segmental vessels were ligated unilaterally at seven levels across the apex of the scoliosis. This patient was treated early in our series, when intra-operative spinal cord monitoring was not available.

Following the anterior surgery, there was a flaccid paralysis of the right leg. The second stage was delayed to allow possible neurological recovery, but this did not occur. After one month she underwent posterior spinal fusion with instrumentation to stabilise the spine, but with no attempt at further correction. There were no additional neurological complications.

Spinal cord monitoring with the use of either somatosensory evoked potentials or combined somatosensory evoked potentials and motor evoked potentials was performed intra-operatively in 331 anterior spinal procedures, including the remaining 28 patients with 'a spinal cord at risk'. None of these patients underwent pre-operative spinal cord monitoring, and baseline traces were obtained when the patient was positioned on the operating table and before the administration of neuromuscular blockade. The intra-operative spinal cord monitoring showed no evidence of reductions in the recorded amplitude of the somatosensory evoked potentials, change in the latency of the somatosensory evoked potentials, or

loss of the motor evoked potentials in any patient before or after ligation of the segmental vessels.

Discussion

Anterior approaches to the spine with or without instrumentation are occasionally recommended in the surgical treatment of severe types of spinal deformity, either in isolation or followed by a posterior spinal fusion.

Neurological complications can occur following corrective anterior spinal surgery and may be due to vascular or mechanical causes. Leung et al⁶ reviewed 871 patients who underwent anterior spinal surgery and reported five (0.6%) with significant post-operative neurological deficits. In a recent report by the Scoliosis Research Society Morbidity and Mortality Committee⁷ the neurological complication rates for all causes following anterior and combined anterior/posterior spinal procedures to correct an adolescent idiopathic scoliosis were 0.26% and 1.75%, respectively.

The anterior procedure usually requires ligation of the segmental vessels on the convex side of the vertebral bodies to provide access to the body and intervertebral disc. As the segmental vessels are part of the complex vasculature of the spinal cord, paraplegia with motor and sensory loss and incontinence could occur simply by ligating these vessels. This risk is reported to range between 0% and 0.86%.^{8,9} The risk factors that have been suggested to potentially impair the blood supply of the spinal cord include segmental vessel ligation at multiple or bilateral levels, a kyphotic deformity, a neoplastic lesion or spinal dysraphism, a left surgical approach, hypotension induced during surgery, partial or complete vertebral column resection, previous anterior spinal surgery and increased patient age.^{6,9-14}

Our only post-operative neurological deficit, which affected one leg, occurred following an anterior convex vertebral column resection and concave spinal osteotomy with unilateral ligation of the segmental vessels performed through a left thoracic approach in a patient with a congenital thoracic scoliosis measuring 127° associated with a diastematomyelia at the level of the congenital vertebral abnormalities. In this patient, the congenital anomalies were in the lower thoracic spine at the critical vascular zone of the spinal cord as defined by Domisse.¹ We believe this neurological complication occurred as a consequence of ischaemic damage to one stem of the diastematomyelia in the presence of a congenitally abnormal blood supply.

Winter et al⁸ reviewed 533 paediatric and adolescent patients who underwent anterior procedures in the thoracic or thoracolumbar spine and reported no neurological complications. They concluded that there is no risk of paraplegia, provided the segmental vessel ligation is unilateral, performed at mid-vertebral body level on the convexity of the scoliosis and without hypotensive anaesthesia.⁸ In our series, hypotensive anaesthesia was used electively in all patients.

In contrast, Bridwell et al⁹ reported that four (1.1%) of 349 patients developed a neurological deficit after anterior

and posterior corrective spinal surgery. In three of these the neurological deficit was attributed purely to a vascular aetiology. In the fourth a combined vascular and mechanical aetiology was suggested. This study concluded that patients who undergo combined anterior and posterior spinal surgery for hyperkyphosis are at the greatest risk for developing neurological complications, and that this is due primarily to anterior interruption of the blood supply to the spinal cord.⁹

Mirovsky et al¹⁵ reported 29 patients who underwent anterior instrumented thoracolumbar fusion and were able to spare the segmental vessels in seven patients, in whom a single rod construct with transvertebral screws 6.25 mm to 7 mm wide were used. There were no neurological complications. In our series, 112 patients had anterior spinal instrumentation with unilateral ligation of the segmental vessels, also without neurological problems.

Bassett, Johnsen and Stanley¹⁶ reported 16 patients who underwent selective spinal angiography followed by anterior spinal surgery with temporary clamping of the segmental arteries and somatosensory evoked potentials monitoring. They recorded no loss of somatosensory evoked potentials after temporary unilateral occlusion of the segmental vessels, and postulated that there was sufficient perimedullary collateral circulation in the spinal cord to allow the routine unilateral ligation of the segmental arteries without increasing the risk of neurological damage.

Apel et al¹⁷ performed clamping of the segmental arteries and monitored somatosensory evoked potentials in 44 patients who underwent anterior spinal fusion to correct thoracic or thoracolumbar spinal deformities from various causes. They noted seven patients in whom there was an alteration in somatosensory evoked potentials after clamping the segmental vessels at or within one vertebral level of the apex of the curve. This reversed once the occlusion was released. From these findings, they recommended temporary segmental arterial occlusion with somatosensory evoked potentials monitoring during anterior thoracolumbar spinal fusion, especially in patients with a congenital kyphoscoliosis. None of our patients had temporary clamping of the segmental vessels before they were divided.

Leung et al⁶ monitored 871 patients who underwent elective anterior spinal deformity surgery with the use of intra-operative somatosensory evoked potentials. The incidence of significant somatosensory evoked potentials changes, as well as post-operative paresis, was much higher in patients who had associated intraspinal anomalies. The authors concluded that patients with cord abnormalities should undergo spinal cord monitoring with soft clamping of the segmental vessels during anterior spinal deformity surgery.

In our series, the patient who developed a neurological deficit after ligation of the segmental vessels was operated

on prior to 1985, when spinal cord monitoring was not available. If this had been available, we believe it would have been helpful, based on the severity of the congenital vertebral and intraspinal anomalies and the extent of the surgery. Also, temporary occlusion of the segmental vessels combined with recording of somatosensory evoked potentials and motor evoked potential signals might have detected impending neurological injury. Pre-operative nerve conduction studies and somatosensory evoked potentials recording could also be helpful in establishing baseline function prior to spinal surgery in patients with intraspinal anomalies or abnormal neurological findings.

In conclusion, this review of 346 consecutive patients who had an anterior spinal procedure for deformity correction revealed only one case of neurological deficit related to segmental vessel ligation. In our experience, segmental vessel ligation appears to carry no risk of causing neurological compromise, unless performed in patients with complex congenital spinal deformities occurring primarily in the thoracic spine, especially those associated with spinal dysraphism at the same level. In this group of patients, intra-operative spinal cord monitoring with the use of somatosensory evoked potentials and preferably motor evoked potentials, as well as soft clamping of the segmental vessels before division, could possibly detect impending neurological injury at a stage when it is reversible.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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Section F

Neuromuscular Deformities

ANTERIOR AND POSTERIOR INSTRUMENTATION AND FUSION OF THORACOLUMBAR SCOLIOSIS DUE TO MYELOMENINGOCELE

MICHAEL J. McMASTER

From the Edinburgh Scoliosis Unit

Twenty-three patients with severe paralytic thoracolumbar scoliosis due to a myelomeningocele were treated by a two-stage procedure. Before operation the mean scoliosis was 98°; after the first-stage procedure, an anterior spinal fusion and correction with Dwyer instrumentation, this was reduced to a mean of 45°. Approximately two weeks later a posterior spinal fusion with Harrington instrumentation was performed, further reducing the scoliosis to a mean of 29°. The pelvic obliquity also was reduced from a mean of 32° to 6°. Although such management carries risks (one patient died of cardiorespiratory failure after the first stage and one patient was made worse), 21 of the 23 patients had improved posture and function.

Children with a lumbar myelomeningocele who have total paralysis or extensive weakness of their spinal musculature may develop a very severe and characteristic scoliosis (Shurtleff et al. 1976; Pigott 1980). At birth the spine is straight but, as the child grows and assumes an upright position, the spine collapses due to gravity; it then continues to deteriorate until the rib cage abuts onto the pelvic rim. The typical deformity is a long thoracolumbar lordoscoliosis extending from the upper thoracic region down to the pelvis and associated with pelvic obliquity (Fig. 1). The child sits on one buttock, needing his arms for support (Fig. 2), and pressure sores may develop in the anaesthetic skin beneath the ischium. Brace therapy may slow the deterioration but is never effective in the long term, and the only treatment for severe progressive curves is to correct and stabilise the spine by operation. Unfortunately, neither anterior nor posterior fusion alone has produced satisfactory results (Sriram, Bobechko and Hall 1972; Baker and Sharrard 1973). More favourable results have been obtained in the few reported series where anterior and posterior spinal fusion with instrumentation are combined, although there is still a high incidence of complications (Brown 1978; Mayfield 1981; Osebold et al. 1982).

This paper reports the results of combined anterior and posterior spinal instrumentation and fusion using

supplementary homologous bone grafts and early mobilisation in a removable underarm plastic jacket.

PATIENTS AND METHODS

From 1976 to 1984, 23 consecutive patients with severe paralytic thoracolumbar scoliosis due to a myelomeningocele were admitted to the Edinburgh Scoliosis Unit. The method of treatment was by anterior spinal fusion with Dwyer instrumentation, later followed by a posterior fusion with Harrington instrumentation. Twenty-two patients had both stages of treatment (one patient having died of cardiorespiratory failure after the first stage) and all operations were performed by the author. There were 14 girls and nine boys whose mean age at operation was 12 years 5 months (range 9 years 9 months to 17 years 6 months). Twenty-one of the children were confined to a wheelchair most of the time because of the severity of their neurological defect; the remaining two patients were not as severely affected and could walk with the aid of calipers and sticks or crutches.

Indications for operation. Any patient over the age of 10 years with severe progressive scoliosis was considered to be a candidate for operation; however, the development of a loss of function was considered to be more important than the actual degree of the curvature. The main indication in all 23 patients was a loss of balance while sitting which required the use of the arms for support (Fig. 2). Five patients also had recurrent pressure sores under one buttock, and three complained of backache.

Eight of the 23 children with progressive curves requiring treatment were seen before the age of 10 years. Operation for these young children was delayed and, instead, a polyethylene total contact thoracolumbosacral

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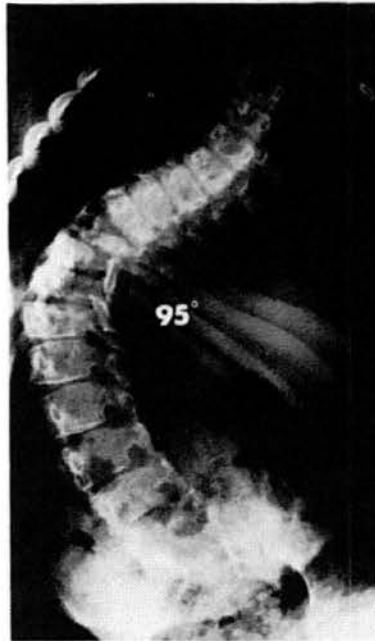


Fig. 1

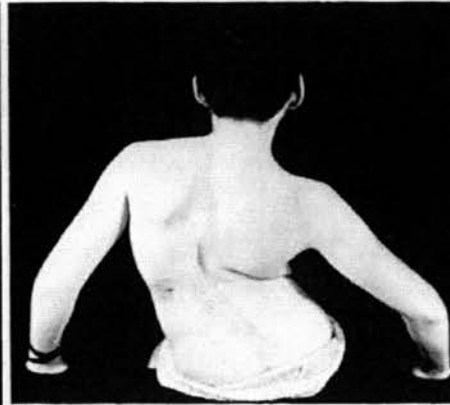


Fig. 2

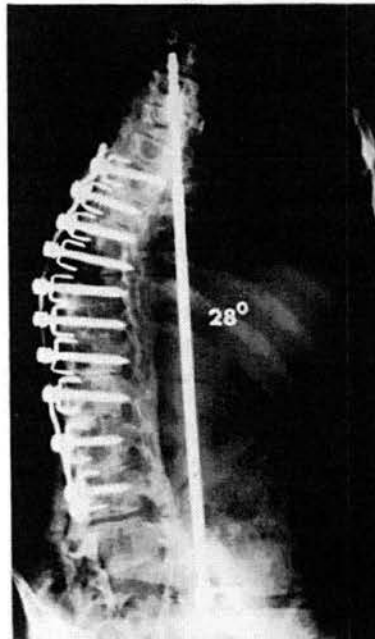


Fig. 3

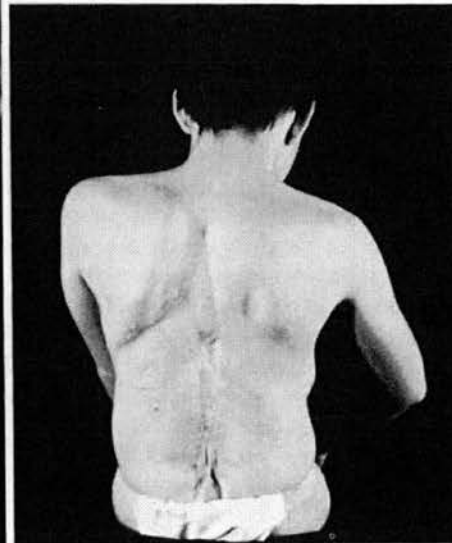


Fig. 4

Case 2. Figure 1 - A boy aged 11 years with a thoracolumbar scoliosis of 95° and a pelvic obliquity of 28°. Figure 2 - The patient was unable to sit upright and had to use his arms for support. There was a list to the left and all his weight was taken on his left buttock. Figure 3 - Four years and three months after a two-stage operation and fusion, good correction has been maintained and there is solid fusion. The scoliosis was 28° and the pelvic obliquity 5°. Figure 4 - He was able to sit well balanced without having to use his arms for support. His head was directly over a level pelvis and his weight equally distributed on both buttocks.

orthosis was worn until a mean age of 10 years 1 month to permit adequate longitudinal spinal growth and sufficient bone stock for a more successful fusion. The optimum time for spinal surgery is between the ages of 10 and 12 years, before the onset of the adolescent growth spurt when these curves deteriorate much more rapidly and cannot be controlled by conservative means. Unfortunately, nine patients were not seen until after the age of 13 years when they already had severe rigid curves which were progressing rapidly.

The fusion was planned to extend over the entire length of the spinal deformity as seen in both the anteroposterior and lateral radiographs, with the upper level two or three vertebrae above the end vertebra, and the lower level at the sacrum.

Operative technique. The correction and fusion of the spine was carried out in two operative stages; the interval between these stages was a mean of 16 days (range 14 to 21 days).

Anterior fusion with Dwyer instrumentation. The

intervertebral discs and the end-plates of the vertebrae spanning the central part of the curve were excised (range 5 to 11 vertebrae, mean 7.5). The disc spaces were then packed with bone chips from an excised rib and correction was obtained by applying Dwyer instrumentation to all the fused levels.

Posterior fusion with Harrington instrumentation. Further correction was obtained by means of a long square-ended Harrington distraction rod applied posteriorly from the upper end of the deformity to the ala of the sacrum on the high side of the pelvic obliquity (see Figs 3 and 6). This rod was pre-bent in the sagittal plane to accommodate the thoracic kyphosis and lumbar lordosis (see Fig. 7). In 18 patients the site of the upper hook was reinforced with methylmethacrylate cement to prevent the hook from cutting through the osteoporotic bone often present in these patients. In the five most recent patients, the distraction rod was further stabilised by securing it with segmental sublaminar wiring to the normal vertebrae in the upper part of the fusion area (Figs 5 to 7).

From 9 to 15 vertebrae (mean 12.5) were fused posteriorly and in all cases this included the sacrum. In 21 patients there was insufficient donor bone graft in the iliac crests, and one or two deep-frozen degenerate femoral heads (removed during total hip replacements) were used as a source of homologous bone grafts to supplement the fusion. An interfacetal intertransverse fusion (Moe 1972) was performed in the upper part of the fusion area where there were normal posterior bony elements. However, because fusion was much more difficult in the lumbar region at the site of the spina bifida defect, large amounts of bone graft material cut into strips were laid in the paraspinal gutters and on the sides of the defective laminae as far as the tips of the vestigial transverse processes, from the lower thoracic region to the ala of the sacrum on both sides. If possible, the patient's own iliac bone was used at the lumbosacral junction.

As prophylaxis against infection, cephradine was given intramuscularly with the premedication, as an intravenous bolus during the operation and orally for 24 hours after each operation. The patient was nursed in a normal bed and log-rolled from side to back to side. Approximately 7 to 10 days after the second-stage operation, the patient had usually recovered sufficiently for a plaster mould to be taken of the trunk. From this a bivalved, underarm total-contact polyethylene jacket was made and the patient allowed to sit upright once it was fitted (Fig. 8). This jacket was light and comfortable, and allowed regular inspection for pressure sores. The jacket was only worn while the patient was upright and removed while in bed. The mean period in hospital for the 22 patients was 8.6 weeks (range 4 to 15 weeks) and six of these patients were only detained for prolonged periods for social reasons. The jacket was worn for a mean of 10 months (range 9 to 12 months) from the time of operation.

RESULTS

Twenty-two patients were followed for a mean of 3 years 9 months (range 1 year 3 months to 7 years 3 months) from the time of the second-stage procedure; 16 patients reached skeletal maturity. One patient died after the first-stage operation. Other details and complications are shown in Table I.

Before treatment the mean scoliosis was 98° (range 65° to 152°); after the first-stage correction with Dwyer instrumentation this was reduced to a mean of 45° (range 4° to 92°) in all 23 patients. In the 22 patients treated by the second-stage correction with Harrington instrumentation, the curve was further reduced to a mean of 29° (range 0° to 62°); at final follow-up, the correction had deteriorated slightly to a mean of 36° (range 12° to 64°).

Pelvic obliquity before operation was a mean of 32° (range 7° to 70°); after the two-stage correction this was reduced to a mean of 6° (range 0° to 20°) and at final follow-up was a mean of 9° (range 0° to 21°).

Before operation five patients had pressure sores under one ischium from sitting with their weight on one buttock; after operation all of these sores had healed but had later recurred in two patients. Twenty-two patients had listed to one side before operation and had needed one or both hands for support while sitting (see Fig. 2). After the two-stage correction, however, 19 had regained their balance and did not require their arms for support (see Fig. 4); the remaining three patients had improved, but still listed to one side by more than 3 cm and required one arm for support.

All but one of the patients had better mobility after the two-stage operation. The condition of 20 patients who had been confined to a wheelchair was improved because the long stable lever of the spine fused to the sacrum enabled the normal functioning muscles in the upper body to have more control over the pelvis and lower limbs, making it easier to transfer to and from the wheelchair. One of the patients who walked regularly with long-leg calipers and crutches improved from a "swing to" to a "swing through" gait which was more economical.

Only one patient (Case 3) was made worse by the operation: this was a girl aged 10 years 8 months who had been able to walk with the aid of sticks and calipers but found that, after operation, she was unable to walk and required crutches for support. There was no worsening of her neurological deficit. Before operation she had had bilateral hip flexion contractures of 45° but had been able to compensate for this by standing with a greatly increased lumbar lordosis; after treatment, however, the lumbar spine was flattened (causing her to be thrown forward at the hips) and she was unable to maintain her balance without using crutches. This problem did not become fully apparent until four weeks after the second-stage operation and was not helped by removal of the Harrington rod. Extension osteotomies of the hips were

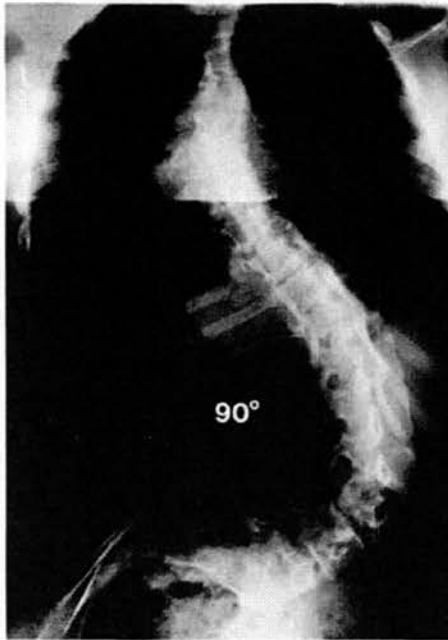


Fig. 5

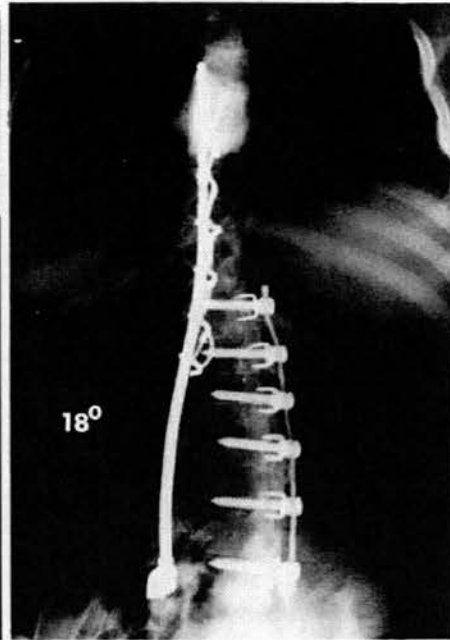


Fig. 6



Fig. 7

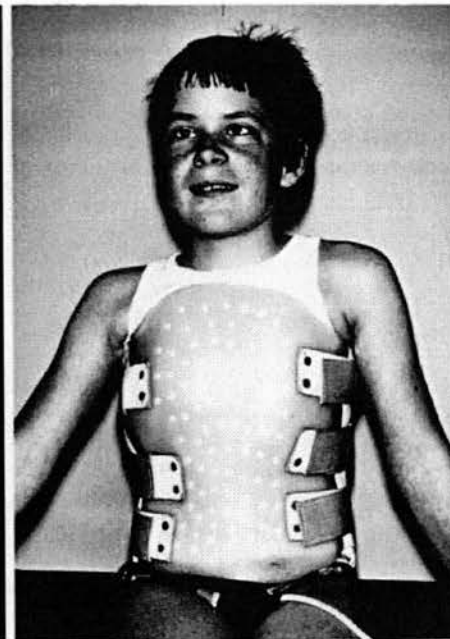


Fig. 8

Case 20. Figure 5 - A girl aged 10 years with a thoracolumbar scoliosis of 90° and a pelvic obliquity of 40°. Figure 6 - Two years after a two-stage correction there is solid fusion. The scoliosis has been reduced to 18° and the pelvic obliquity to 7°. The Harrington distraction rod was stabilised by segmental sublaminar wiring and the site of the upper hook reinforced with methylmethacrylate cement. Figure 7 - The normal lumbar lordosis has been maintained by bending the distraction rod before operation. Figure 8 - The bivalved underarm total-contact polyethylene jacket was custom-made and worn in the postoperative period.

Table I. Details of 23 myelomeningocele patients with paralytic scoliosis treated by the two-stage procedure

Case	Extent of fusion	Fused vertebrae		Homologous bone graft	Bone cement	Segmental sublaminae wiring	Follow-up		Scoliosis			Pelvic obliquity				Complications
		Anterior	Posterior				(years)	(months)	Preop.	Postop. (Dwyer)	Postop. (Harrington)	Final	Preop.	Post-op.	Final	
1	T4 S1	8	15	Yes	No	No	4	1	120°	78°	48°	54°	58°	6°	10°	None
2	T5 S1	10	14	Yes	Yes	No	4	6	95°	43°	28°	30°	28°	0°	5°	Erosion of bone by lower Harrington hook; minor postop. wound necrosis
3	T9 S1	5	9	Yes	No	No	5	0	83°	40°	30°	42°	7°	0°	0°	Unable to walk with calipers after operation
4	T4 S1	9	15	Yes	No	No	6	3	76°	42°	20°	36°	35°	0°	0°	Postop. wound infection healed after 5 weeks
5	T6 S1	11	13	Yes	No	No	4	2	152°	62°	36°	49°	70°	20°	21°	Erosion of bone by lower Harrington hook
6	T6 S1	11	13	Yes	Yes	No	4	4	78°	36°	26°	38°	20°	6°	8°	Minor postop. wound infection; lumbosacral pseudarthrosis
7	T7 S1	10	12	Yes	No	No	7	3	83°	9°	0°	15°	26°	6°	13°	None
8	T7 L5	10	Not done	No	No	No			143°	66°	-					Died of cardiorespiratory failure after first stage
9	T7 S1	9	12	Yes	Yes	No	6	0	126°	92°	60°	62°	22°	10°	10°	None
10	T7 S1	5	12	Yes	No	No	3	6	100°	45°	22°	26°	23°	8°	8°	None
11	T6 S1	7	13	Yes	Yes	No	5	0	102°	49°	33°	40°	40°	8°	19°	Posterior wound necrosis (required excision and resuturing)
12	T7 S1	7	12	Yes	Yes	No	3	0	106°	50°	36°	39°	35°	9°	10°	None
13	T8 S1	5	11	Yes	No	No	4	0	90°	59°	35°	49°	56°	10°	12°	None
14	T8 S1	6	11	Yes	No	No	4	0	110°	53°	36°	45°	18°	0°	3°	None
15	T4 S1	9	15	Yes	Yes	No	4	0	108°	58°	42°	52°	38°	0°	8°	Minor postop. wound necrosis
16	T6 S1	8	13	Yes	Yes	No	3	3	90°	38°	28°	33°	34°	10°	12°	None
17	T8 S1	6	11	Yes	Yes	No	2	0	86°	39°	30°	34°	12°	3°	3°	None
18	T8 S1	6	11	Yes	Yes	No	4	0	65°	4°	0°	12°	17°	0°	0°	None
19	T9 S1	6	10	Yes	Yes	Yes	2	6	78°	41°	30°	36°	43°	8°	8°	Sacral pressure sore under jacket
20	T5 S1	7	14	Yes	Yes	Yes	2	1	90°	23°	18°	20°	40°	7°	7°	Minor postop. wound necrosis
21	T6 S1	6	13	Yes	Yes	Yes	2	0	85°	40°	30°	18°	30°	14°	14°	None
22	T5 S1	9	14	Yes	No	Yes	1	6	90°	70°	62°	64°	26°	15°	15°	None
23	T6 S1	7	13	Yes	No	Yes	1	3	96°	10°	3°	7°	35°	0°	5°	None

performed one year later and, although this helped, it did not fully restore her walking ability.

Complications. No patient had increased neurological deficit, but other complications occurred in eight patients.

Death. A girl aged 9 years 11 months (Case 8) died of cardiorespiratory failure. Before operation she had had an extremely severe thoracolumbar scoliosis of 143° extending from T5 to the sacrum, and she already had impaired respiratory function. After a very extensive anterior fusion with Dwyer instrumentation involving 10 vertebrae from T7 to L5, her scoliosis was reduced to 66°. Initially she appeared to be recovering well but then developed cardiorespiratory failure and died 18 hours after operation.

Wound necrosis. Four patients (Cases 2, 11, 15 and 20) developed varying degrees of necrosis affecting the margins of the wound in the scarred lumbar region at the site of the closure of their myelomeningocele. In two patients, the wound necrosis was minor and required no treatment. In one patient with more severe necrosis, an operation was required to excise the wound edges and perform secondary closure; this wound then healed.

Wound infection. Two patients (Cases 4 and 6) developed infections of their posterior wound. Both were treated with antibiotics; one infection resolved rapidly over a period of a few days and the other continued to drain for five weeks before healing. There were no further problems.

Pressure sores. One patient (Case 19) developed a

pressure sore due to the rubbing of his polyethylene jacket over his sacrum. This healed after temporary removal of the jacket and two weeks of bed rest.

Instrument failure. In no patient did the implanted instrumentation either break or become loose before the spine had fused. However, in two patients (Cases 2 and 5) there was evidence of bony erosion where the lower hook had been applied to the alae of the sacrum; this resulted in a mild loss of correction over the first nine months (8° in one patient and 13° in the other), but subsequently the spine in both became stable as the fusion consolidated.

Pseudarthrosis. Only one patient (Case 6) developed what appeared to be a pseudarthrosis at the lumbosacral junction which resulted in a 12° loss of correction. No treatment was required and the spine became stable after 18 months. At final follow-up after four years four months, there was no further loss of correction and the pseudarthrosis had apparently healed.

DISCUSSION

Severe unbalanced thoracolumbar scoliosis and pelvic obliquity of some patients with a myelomeningocele present a challenging problem; the muscle paralysis and sensory deficit make operative management much more difficult than that of other patients with scoliosis. The aim of operation should be to correct the deformity and to fuse the spine so that the trunk is supported in a balanced position over a stable and level pelvis. This should allow the patient to have independent use of the arms for activities other than supporting the trunk and should reduce the incidence of pressure sores under the buttocks. The ability to walk in long-leg calipers may be improved by providing a more stable lever (of the spine fused to the sacrum) to control the pelvis. In this series, one or more of these objectives were achieved in 18 of the 22 patients (82%). A further three patients with very severe deformities were improved, although they did not achieve complete balance of the trunk.

This type of spinal deformity requires both anterior and posterior surgery for several reasons. Firstly, the scoliosis is very rigid and, in order to gain as much correction as possible, extensive release and instrumentation of the spine from both sides is necessary. The mean curvature of the scoliosis in the 23 patients before operation was 98° ; this was improved by a mean of 54% after the anterior operation and further improved to 70% after the posterior operation. The pelvic obliquity was reduced from a mean of 32° to 6° , a correction of 80%. The addition of methylmethacrylate cement to reinforce the osteoporotic bone at the site of the upper Harrington hook and segmental sublaminar wiring around the distraction rod were useful aids in stabilising the posterior instrumentation. These results compare favourably with those of Osebold et al. (1982) who reported 17 myelomeningocele patients with a mean pre-operative scoliosis of 81° which was improved by a mean

of 62% after the same two-stage procedure.

Secondly, these are very long curves, extending from the upper thoracic region down to the pelvis; in order to stabilise the spine and control the pelvic obliquity, it is essential to fuse the full extent of the deformity. With anterior operation alone, the inclusion of both the upper thoracic region as well as the sacrum is technically difficult but this can be done by means of a second-stage posterior fusion (Figs 3 and 6)

Thirdly, posterior fusion with Harrington instrumentation when used alone may fail because the large spina bifida defect and hypoplastic posterior bony elements provide an inadequate bed for bone grafting; this may result in an incidence of pseudarthrosis as high as 46% (Sriram et al. 1972; Osebold et al. 1982). However, using the combined technique, the thoracolumbar spine is spanned both anteriorly and posteriorly, thus greatly reducing the incidence of pseudarthrosis. In this series only one patient (4%) developed a pseudarthrosis at the lumbosacral junction. There were no complications related to the use of degenerate femoral heads as homologous bone grafts to supplement the posterior fusion; Osebold et al. (1982) used the same technique and reported a 23% incidence of pseudarthrosis, most of which occurred at the lumbosacral junction.

Although the combined technique provides an excellent means of correcting and stabilising the spine, it is not without its serious complications, a fact attested to by the death of one patient after the first-stage thoracotomy, and the impaired mobility of a second patient who could walk with sticks and calipers before the operation but was unable to do so afterwards.

In conclusion, the operative management of this type of spinal deformity is an arduous and complex task, but it can be of great benefit to these severely disabled children and their families. There is, however, a high potential for disaster and great care as well as extensive experience is required.

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The Long-Term Results of Kyphectomy and Spinal Stabilization in Children with Myelomeningocele

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Ten myelomeningocele patients with a severe lumbar kyphosis were treated by resection of their kyphus, internal fixation, and spinal fusion. A mean kyphosis of 131° was reduced to 44° following the surgery. This was a very major procedure, associated with many complications. One patient died during the surgery and the remaining nine patients were followed for a mean of 7 years 4 months to skeletal maturity. The most successful methods of internal fixation were either by two Harrington distraction rods combined with compression across the osteotomy site (Group 3) or by a posteriorly applied AO plate (Group 2). A long posterior fusion extending from the mid-thoracic region to the sacrum was necessary to provide long-term stability and prevent the development of a thoracic lordosis. At skeletal maturity, all seven patients in Groups 2 and 3 had a flat back without pressure sores and all were able to sit upright without using their arms for support. [Key words: myelomeningocele, kyphosis, kyphectomy, skeletal maturity]

A VERY CHARACTERISTIC angular kyphosis may be present at birth in a small minority of patients with a thoracolumbar myelomeningocele and a severe neurological deficit. This kyphosis typically occurs in the lumbar region with its apex at the second or third lumbar vertebra. The associated spina bifida is very wide and as a result the paraspinal muscles, which normally extend the spine, are displaced forward and act at a mechanical disadvantage.^{1,5} Once the child begins to sit, the kyphosis rapidly deteriorates, becoming very severe and rigid at an early age. Sharrard and Drennan (1972)¹² in a study of 35 neonates found that the kyphosis frequently exceeded 80° at birth and by 8 years the majority of curves were over 170° . A compensatory thoracolumbar lordosis develops above the kyphosis and this further accentuates the very angular gibbus. The upper body is thrown forward and the child is forced to prop himself up with both arms (see figures 3, 8, 13). Progression is stopped only when the rib cage comes to rest on the front of the thighs. Sitting or lying is difficult and the anaesthetic-scarred skin over the kyphosis easily develops pressure sores. Respiration may be impaired by the upward pressure of the compressed abdominal contents against the diaphragm.

In an attempt to prevent these problems from developing, Sharrard (1968)¹¹ introduced his technique of spinal osteotomy and resection of the kyphosis in the newborn. Unfortunately, he found that the deformity tended to recur and later extended the technique to older children with more successful results.¹² Since this time other surgeons have used similar techniques and found this to be a very major procedure associated with many complications, including

death^{2-4,6-9,13}. However, in none of these series has there been a long-term review of the effectiveness of the procedure in preventing recurrence of the deformity.

This paper reports the long-term results of kyphectomy and stabilization in a series of patients followed to skeletal maturity. The objective of surgery was to produce a flat back without any bony prominences likely to cause pressure sores and to stabilize the spine sufficiently to allow the patient to sit upright and have independent use of the arms.

MATERIALS AND METHODS

From 1976 to 1980, ten consecutive patients with a severe lumbar kyphosis due to a myelomeningocele were treated at the Princess Margaret Rose Hospital, Edinburgh, by resection of their kyphosis, internal fixation, and spinal fusion (Table 1). All of the operations were performed by the author. There were six boys and four girls whose mean age at operation was 10 years (range 8 years to 12 years 7 months). The mean kyphosis prior to surgery was 131° (range 115° to 166°). All of the patients had normal function in their upper limbs; but from the waist down there was a severe neurological deficit with no motor or sensory function. All were confined to a wheelchair and were unable to wear long leg calipers because the pelvic band rubbed on the kyphosis.

Indications for Surgery. All of the patients had a very severe progressive spinal deformity resulting in a loss of sitting balance and necessitating the use of the arms for support. Four patients had recurrent pressure sores over the apex of their kyphosis.

These children all sat on the back of their sacrum with their hips fully extended (see Figures 4, 9, 14). However, they all had 90° of passive hip flexion; this is necessary because once the kyphosis is corrected, the pelvis will be realigned and the patients will not be able to sit if the hips cannot be flexed.

Preoperative Preparation. Pressure sores over the kyphosis were treated by debridement and dressings to allow any infection to settle and obtain as much healing as possible. It was not always possible or necessary to obtain complete healing before surgery. All of the patients had a hydrocephalus and the patency of their shunts was assessed by a paediatric neurologist. An IVP was performed to assess kidney function and any urinary tract infection was treated.

OPERATIVE TECHNIQUE

The patient was placed prone on the operating table lying on a specially designed foam block in which a hole had been cut to allow the abdomen to hang free, thus decreasing the intra-abdominal pressure and reducing the bleeding. A midline longitudinal incision was made, regardless of the scarring, over the apex of the kyphosis extending from the mid-thoracic region to the sacrum. If possible any ulcerated area was excised with the skin incision. The skin and subcutaneous tissues, which were usually thin and adherent to the underlying dura, were dissected bilaterally to the margins of the laminar defect, leaving as thick flaps as possible.

Table 1. Details of Ten Myelomeningocele Patients With a Congenital Lumbar Kyphosis Treated Surgically

Case	Sex	Age at operation yr + mths	Neurologic level	Levels of kyphosis	Extent of fusion	Method of internal fixation	Duration of follow-up yr + mths	Kyphosis (degrees)			Complications	Final result
								Pre-op	Post-op	Final		
1	M	11+9	T10	T11-L4 apex L2	T12-L5	Bilateral Dwyer cables posteriorly, T12-L5	7+6	127	70	163	1. Subtrochanteric fracture femur at 6 months 2. Complete loss of correction of kyphosis 3. Severe increase in thoracic lordosis and loss of sitting balance after 2 years 4. Pressure sore and sinus over Dwyer screw head	Very poor
2	M	12+4	T12	T12-L4 apex L2	T12-L5	Bilateral Dwyer cables posteriorly T12-L5	6+0	166	63	93	1. Mid-shaft fracture femur at 1 month 2. Moderate loss of correction of kyphosis 3. Moderate increase in thoracic lordosis 4. Pressure sore and sinus over Dwyer screw head	Poor
3	M	10+9	T10	T11-L4 apex L2	T10-S1	AO plate with 8 screws applied posteriorly T10-L5	6+6	150	14	33	1. One screw partly pulled loose 2. Supracondylar fractured femur at 3 months	Fair
4	F	8+5	T10	T12-L5 apex L2	T10-S1	Bilateral Dwyer cables posteriorly L1-L4 Bilateral Harrington distraction rods T10-S1	8+5	130	32	25	None	Good
5	F	12+7	T10	L1-L5 apex L3	T9-S1	Bilateral Dwyer cables posteriorly T11-L5 Bilateral Harrington distraction rods T9-S1	7+0	115	41	66	1. Supracondylar fracture femur at 6 months 2. One lower Harrington-Hook displaced at 1 yr 3 months but spine remained stable 3. Pressure sore and sinus over Dwyer screw head	Good
6	M	12+7	T12	T11-L4 apex L2 T12-L4 apex L2	T9-S1	Single Dwyer cable posteriorly L1-L4 Bilateral Harrington distraction rods T9-S1	7+9	120			Cardiac arrest and death shortly after spinal cord divided at surgery One Harrington rod broke at 3 years but no loss of correction	Good
7	M	8+8	T11	T12-L4 apex L2	T9-S1	Single Dwyer cable posteriorly L1-L4 Bilateral Harrington distraction rods T9-S1	7+9	132	50	62		
8	M	8+1	T12	T12-L4 apex L2	T8-1	Single Dwyer cable posteriorly L1-L4 Bilateral Harrington distraction rods T8-S1	8+3	125	51	68	Small area wound necrosis	Good
9	F	8+0	T12	T11-L5 apex L2	T7-S1	Bilateral staples Bilateral Harrington distraction rods T7-S1	7+6	120	42	30	1. Small area wound necrosis 2. Supracondylar fracture femur at 8 months	Good
10	F	9+0	T12	L1-L4 apex L2	T7-S1	Bilateral staples posteriorly Bilateral Harrington distraction rods T7-S1	7+6	115	30	12	Lower end of one Harrington rod displaced at 3 years but no loss of correction	Good

The posterior elements were deficient in the midline from the thoracolumbar junction to the sacrum and the residual laminae and pedicles were widely spread with their tips displaced anteriorly. The erector spinae muscles were found to be displaced laterally and anteriorly bowstringing across the sides of the kyphosis. Using sharp dissection, these muscles were stripped from the sides of the kyphosis and the dissection continued to the front of the vertebral bodies where the retroperitoneal tissues were displaced forwards from the apex of the kyphosis. The segmental vessels were ligated at the sides of the vertebral bodies and the nonfunctioning spinal nerves divided as necessary (Figure 1). Fortunately the great vessels do not follow the anterior aspect of the vertebral bodies and bowstring across the kyphosis. At the end of this procedure it was usually possible to pass several fingers anterior to the kyphosis.

Despite the circumferential removal of all of the soft tissues, the kyphosis remained very rigid and correction could only be obtained by resecting several apical vertebrae and shortening the spine. In order to make this easier, the flattened, nonfunctioning spinal cord was dissected free from the vertebral bodies and transected just proximal to the apex of the kyphosis (Figure 2). The dura at the proximal cut end was dissected free and sutured tightly distal to the transected neural elements to allow possible free drainage of CSF out of central canal into the subarachnoid space. The distal part of the dural sac was occasionally resected, but this usually caused extensive bleeding from the epidural veins which was difficult to control.

Using a reciprocating saw, the bony kyphosis was osteotomized, through the middle of the vertebral body just distal to the thoracolumbar junction and through the vertebral body just distal to the apex of the kyphosis. This segment, which usually totalled two to three vertebral bodies, including not only the apex of the kyphosis but also part of the lordotic region just proximal to the apex. Sufficient bone was removed to allow flattening of the spine and close opposition of the everted bone ends, which are bevelled.

Internal Fixation

Correction was maintained by applying posterior instrumentation. The technique of internal fixation evolved with experience, and three different methods were used.

1) Bilateral Dwyer cables (Cases 1 and 2). The two Dwyer cables were applied posteriorly and fixed by screws passing through the

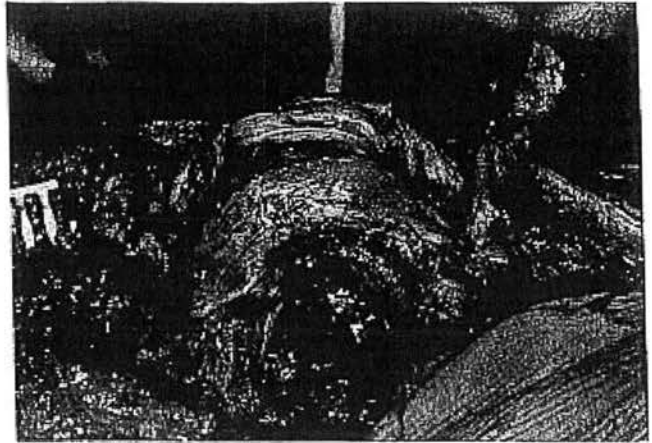


Fig 2. The dissection has been continued to the front of the vertebral bodies and the retroperitoneal tissues displaced forwards from the apex of the kyphosis. Despite circumferential removal of all soft tissues, the kyphosis remains rigid. The nonfunctioning spinal cord is transected to give greater access.

vertebral bodies at two levels above and below the osteotomy site (see Figure 5). The Dwyer cables were in parallel and acted as tension bands to maintain correction and apply compression across the osteotomy. The fixation extended from the thoracolumbar junction to the fifth lumbar vertebrae.

2) AO Compression Plate (Case 3). The plate was applied to the posterior aspect of the vertebral bodies from T10 to L5 (see Figure 10). In order to apply the plate directly to the bone it was necessary to resect the nonfunctioning spinal cord from the thoracolumbar junction to the sacrum; this caused extensive bleeding from the epidural veins which had to be controlled. The plate was fixed by means of eight cancellous bone screws passed through the vertebral bodies, which gave very rigid internal fixation with compression across the osteotomy site.

3) Bilateral Harrington rods supplemented by a single Dwyer cable or staples (Cases 4,5,7,8,9,10). The Harrington rods were applied posteriorly and in parallel from the thoracic region (between T7 and T10) to the sacrum (see Figure 15). Harrington 1253 hooks were inserted under the lamina and into the facet joints in the thoracic region. Moe alar hooks were applied over the ala of the sacrum or driven into the body of the sacrum on either side.

Harrington distraction rods were passed through the thoracic hooks and used not to distract the spine but the lever downward, thus correcting any residual kyphosis, and fitted into the alar hooks bilaterally. This provided a three-point fixation system across the osteotomy site in the same way as Harrington rods have been used to stabilize the spine after a traumatic fracture dislocation. This was supplemented by staples or a single Dwyer cable applied posteriorly to compress the osteotomy site from one level above to one level below.

Spinal Fusion

All of the exposed bony surfaces from the mid-thoracic region to the sacrum were decorticated, including the inside of the spinal canal if possible. Bone grafts from the excised vertebral bodies and, if necessary, homologous bone from deep-frozen degenerate femoral heads (McMaster 1987)¹⁰ were used to perform the spinal fusion. Strips of bone were placed anteriorly across the osteotomy site and a posterior fusion performed from the mid-thoracic region to the sacrum including all of the instrumented levels.

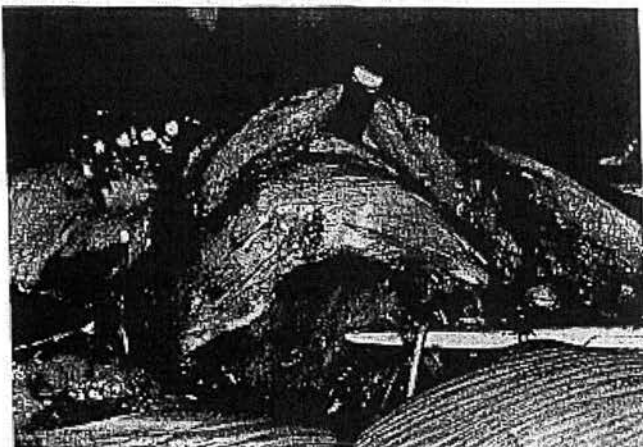


Fig 1. The spine has been exposed posteriorly and the paraspinal muscles stripped from the sides of the kyphosis. The posterior bony elements are deficient in the midline and the nonfunctioning spinal cord is stretched over the apex of the kyphosis. One of the spinal nerves is seen emerging from the side of the kyphosis.

Closure

Once the deformity was corrected, there was usually redundant skin and subcutaneous tissues which allowed excision of any ulcerated areas over the apex of the kyphosis. The paraspinal muscles were pulled posteriorly into their normal position and if possible sutured behind the vertebral column. This provided a soft tissue pad over the instrumentation and any residual kyphosis.

Postoperative Care

As prophylaxis against infection, Cephadrine was given intramuscularly with the premedication, as an intravenous bolus during the operation and orally for 24 hours after the operation. The patients were nursed in a normal bed and log-rolled from side to back to side. After 3 months, a custom-made, bivalved, under-arm, total contact polyethylene jacket was applied and the patient allowed to sit upright. The jacket was worn only while the patient was upright and removed while in bed. Between 12 and 15 months after the surgery, the patient was allowed to sit without support.

RESULTS

One patient (Case 6) died at surgery and the remaining nine patients were followed for a mean of 7 years 4 months (range 6 years to 8 years 5 months) from surgery. At final follow-up the mean age of the nine patients was 17 years 3 months (range 16 years 5 months to 19 years 3 months), and all were skeletally mature.

The patients were treated in three successive groups; each group represented a step in the evolution of the method for internal fixation.

Group 1

These two patients (Cases 1 and 2) were aged 11 and 12 years, and following kyphectomy the spine was stabilized by two Dwyer cables. The fixation and posterior fusion extended from the thoracolumbar junction to the sacrum.

Good initial correction was obtained in both patients. In one patient (Case 1) the kyphosis was corrected from 127° to 70° but after 1 year, when the supporting jacket was removed, the curve deteriorated rapidly and by skeletal maturity measured 163°. In addition, a severe fixed thoracic lordosis developed above the fused area and contributed to the overall deformity. This patient had a very poor result with a loss of sitting balance and recurrent pressure sores developing over the apex of the kyphosis.

The second patient (Case 2) had a better result, with the kyphosis being corrected from 166° to 63° (Figures 3,4,5), and deteriorating to 93° at skeletal maturity (Figure 6). A thoracic lordosis developed but was not severe and at final follow-up the patient was able to sit upright without using his arms for support (Figure 7), and there were no pressure sores.

Group 2

There was only one patient in this group (Case 3): a boy aged 10 years 9 months with a 150° kyphosis (Figures 8,9). Following kyphectomy there was nearly complete correction and this allowed the application of an AO direct compression plate to the posterior aspects of the vertebral bodies from T10 to L5 (Figure 10). In order to apply the plate directly to bone it was necessary to resect the nonfunctioning spinal cord from the thoracolumbar junction to the sacrum. This gave very rigid internal fixation with compression across the osteotomy site, and the spine was fused throughout the instrumented area. After operation the kyphosis was corrected to 14°. At final follow-up at 17 years 3 months, the kyphosis had

deteriorated to 33° and there was a solid fusion (Figure 11). However, the upper screw had partly pulled loose from the vertebra and radiographs showed that a mild thoracic lordosis had developed above the fused area. This, however, was of no functional significance and although the patient had a stunted trunk, he had a flat back without pressure sores and was able to sit well-balanced without having to support himself with his arms (Figure 12).

Although this method of fixation gave a good result, it had the disadvantage of requiring an extensive resection of the dural sac; this caused profuse bleeding from the epidural veins which was difficult to control.

Group 3

In this group of six patients (Cases 4,5,7,8,9,10) an attempt was made to prevent a thoracic lordosis from developing by the posterior application of two Harrington rods in parallel from the thoracic region (between T7 to T10) to the sacrum (Case 7, Figures 13, 14, 15). This was supplemented by a single Dwyer cable or staples applied posteriorly to compress the osteotomy site. The spine was fused posteriorly over the whole of the instrumented area to the sacrum.

Before surgery the mean age of the patients was 9 years 2 months (range 8 years to 12 years 7 months) and their mean kyphosis was 123° (range 115° to 132°). Immediately after surgery the kyphosis was corrected to a mean of 41° (range 30° to 51°). At final follow-up at a mean age of 16 years 8 months (range 15 years 6 months to 19 years 7 months), three patients had improved their correction by a mean of 12° (range 7° to 18°) and three patients had a mean loss of correction of 18° (range 12° to 25°). All had a solid fusion and no patient had a thoracic lordosis. All of the patients had a flat back and there were no pressure sores. All were able to sit erect without using their arms for support (Figures 16 and 17). However, all tended to obesity and had significantly stunted trunks.

COMPLICATIONS

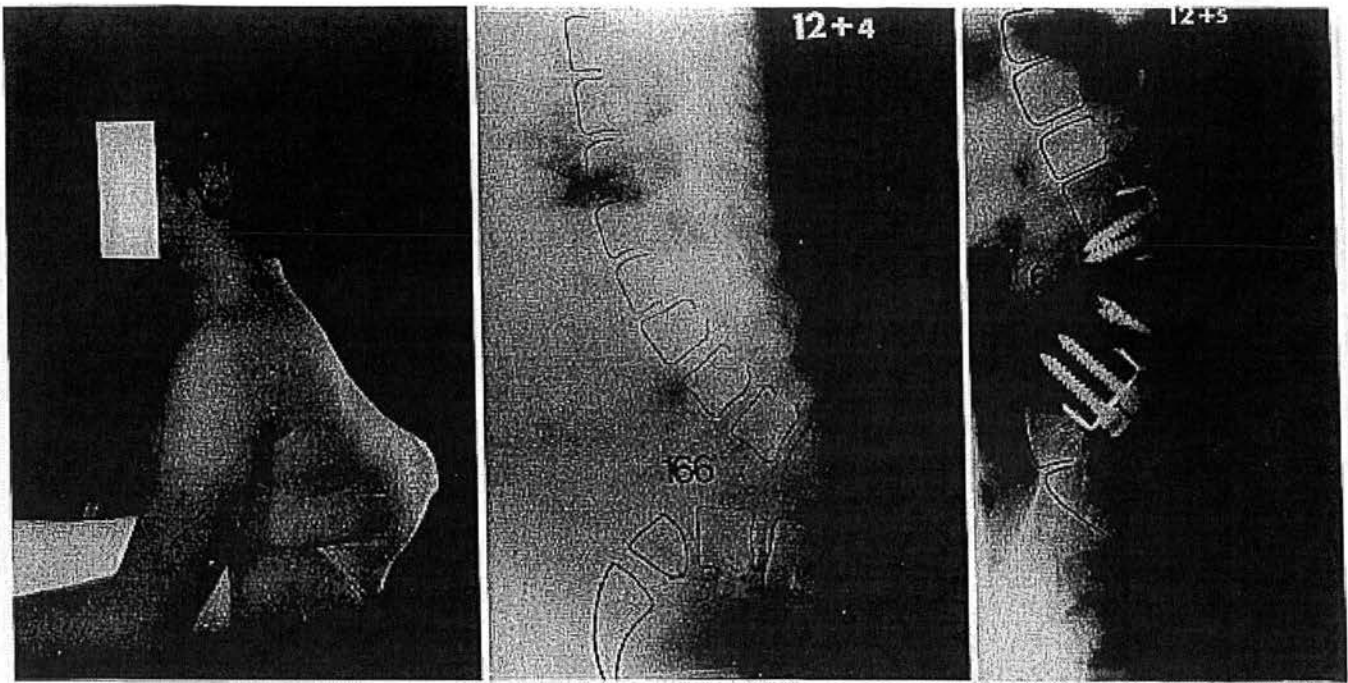
Complications occurred in all but one of the patients (Case 4).

1) Death from a cardiac arrest occurred in one patient (Case 6) shortly after the spinal cord was divided and ligated during the surgery. Winston et al (1977)¹⁴ reported the deaths of two children in similar circumstances and suggested that if the spinal cord and dura are ligated together, it is possible to precipitate an acute elevation of intracranial pressure and coning of the midbrain. They recommended suturing the dura distal to the transected cord to allow drainage of the CSF out of the central canal into the subarachnoid space. This was done in all the subsequent patients without similar complications.

2) A fracture of the femur occurred in five patients at a mean period of 5 months from surgery (range 1 to 8 months). Three of the fractures were supracondylar, one midshaft, and one subtrochanteric. All were due to disuse osteoporosis following the long period of recumbency and occurred with minimal trauma while the patient was being lifted or turned. Two were treated by internal fixation and three in a padded backslint. All healed rapidly and without problem.

3) Instrument failure occurred in four patients. In two patients (Cases 5, 10), one of the lower Harrington hooks displaced and in a third patient the Harrington rod broke (Case 7). In a fourth patient (Case 3) a screw partly pulled loose. In all four patients, the instrumentation failed between 15 months and 3 years after the surgery when the spine was solidly fused and there was minimal, if any, loss of correction (Figures 15,16).

4) A pressure sore and an infected sinus developed over one of



Figs 3-5. (Case 2) **3**, (left), A boy aged 12 years 4 months with a very severe angular lumbar kyphosis. His chest rests on the front of his thighs and he is unable to sit upright without the support of his arms. **4** (middle), The kyphosis measures 166° and he sits on the back of his sacrum with his hips fully extended. **5** (right), Immediately after kyphectomy and internal fixation with two Dwyer cables, the kyphosis was reduced to 63° .

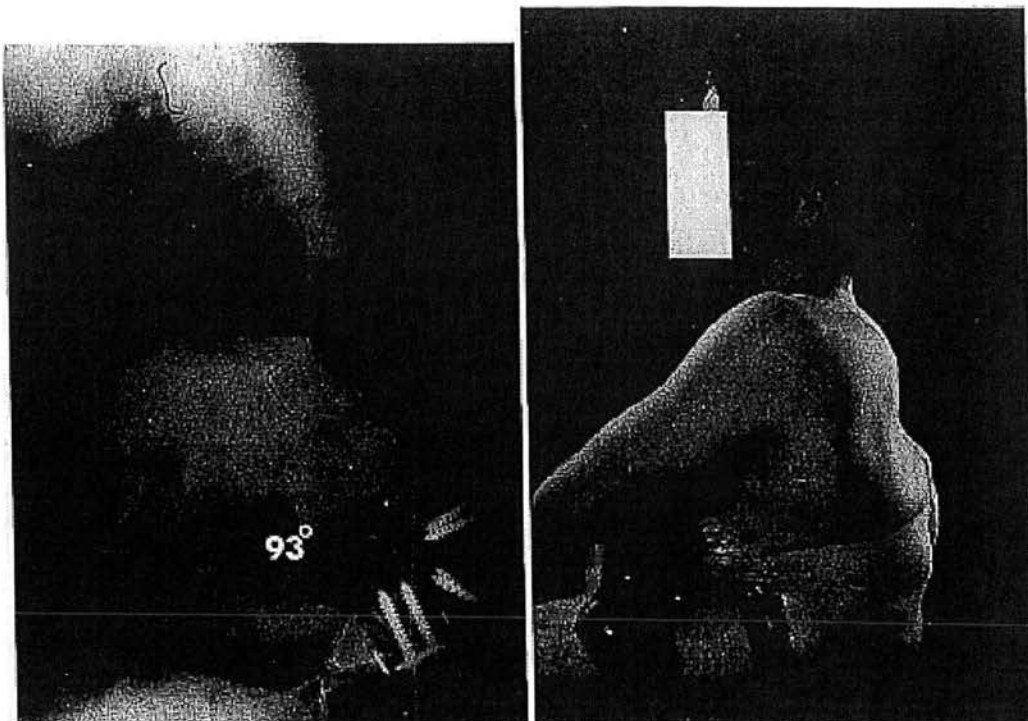
the Dwyer screw heads in three patients (Cases 1,2,5). Healing only occurred after excision of the sinus and removal of the screw.

5) Mild superficial wound necrosis occurred in two patients (Cases 8,9) during the early postoperative period and settled with conservative treatment.

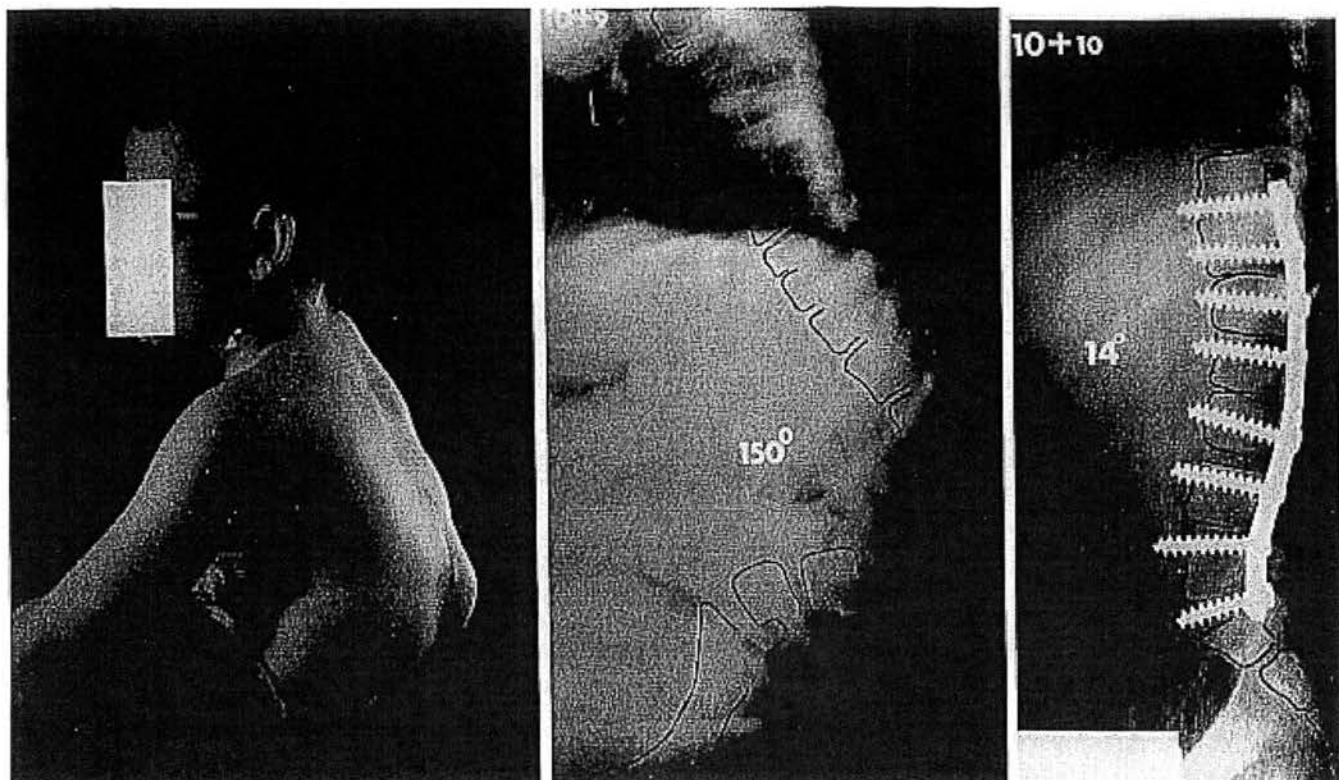
DISCUSSION

A kyphosis presenting at birth in a myelomeningocele child is a very difficult problem because the deformity deteriorates rapidly,

becoming an extreme deformity at an early age¹³ (Figures 3,8,13). These children have a severe neurological deficit and cannot walk. They spend all of their time sitting or lying and this can be difficult because the large angular kyphosis presses against the back of the chair or bed. Brace treatment is ineffective and a potent cause of pressure sores.³ For surgery to be effective, it should produce a flat back without a gibbus likely to cause pressure sores and also provide long-term stability to enable the patient to sit upright in a balanced position without having to use the arms for support.



Figs 6,7. (Case 2) **6** (left), At 17 years 4 months, the kyphosis has relapsed to 93° and a thoracolumbar lordosis has formed above the fused lumbar region. **7** (right), The patient is able to sit upright without using his arms for support.

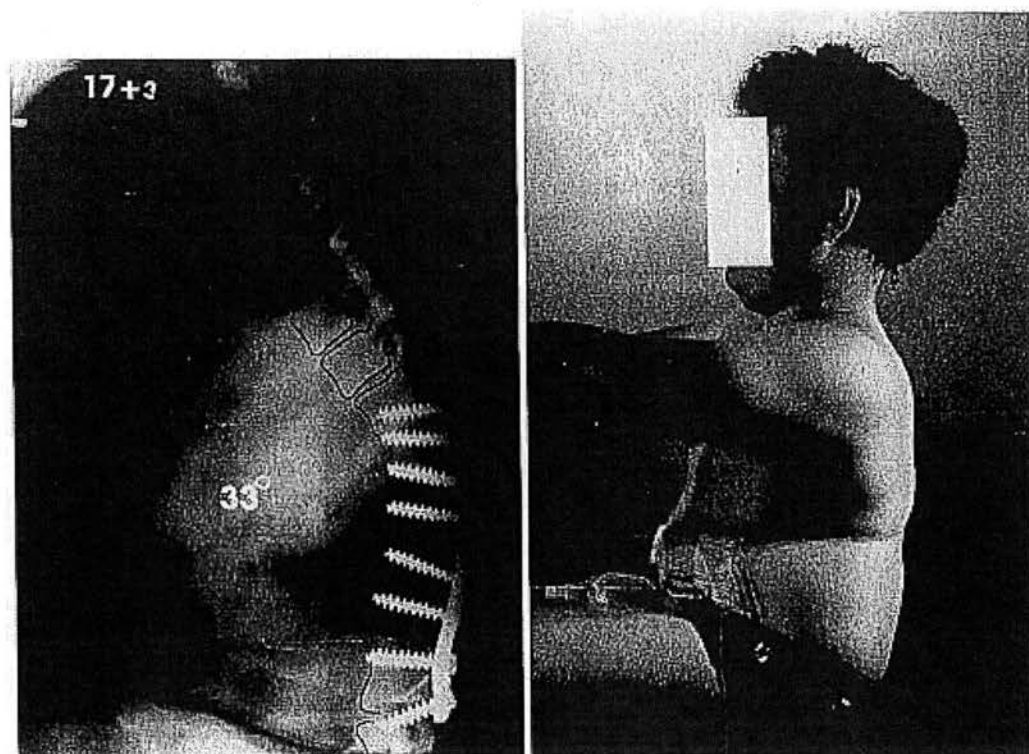


Figs 8-10. (Case 3) **8** (left), A boy aged 10 years 9 months with a severe lumbar kyphosis. **9** (middle), The kyphosis measures 150°. **10** (right), After kyphectomy and internal fixation with an AO compression plate, the kyphosis was reduced to 14°.

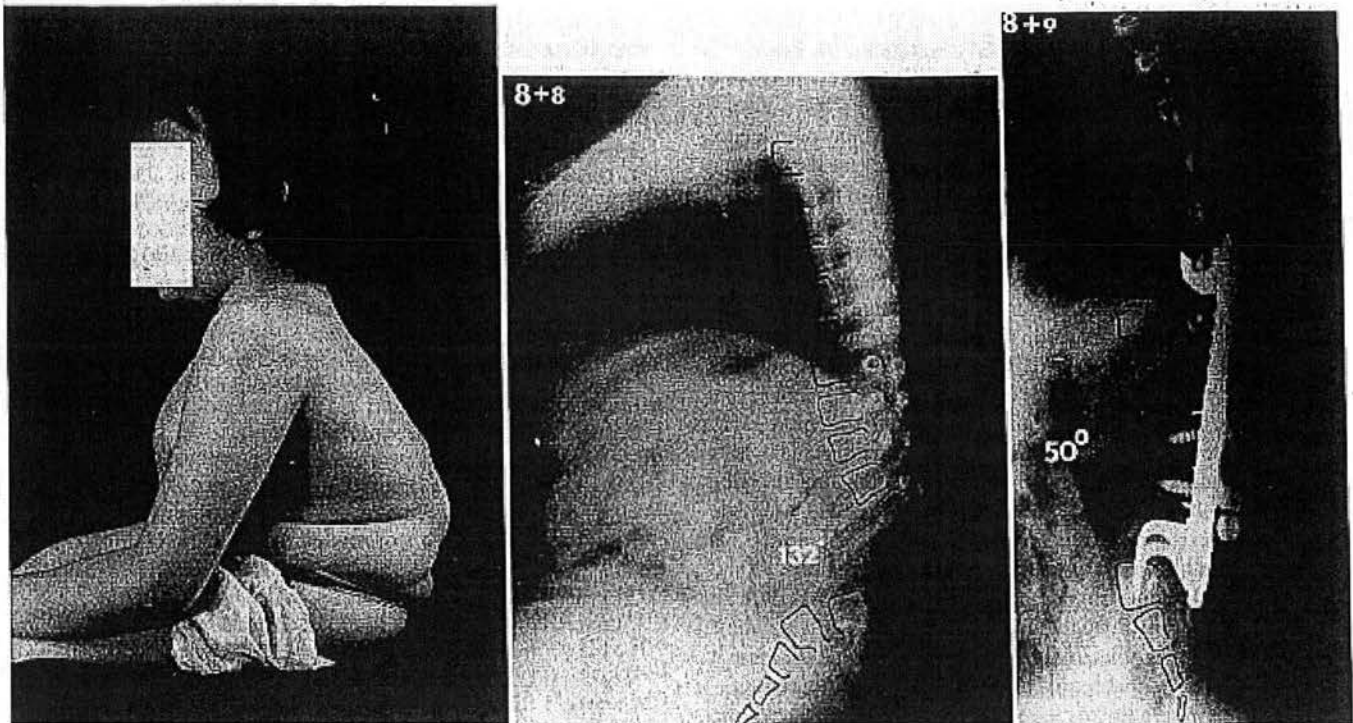
Successful surgical treatment requires excision of the kyphosis to reduce the bony prominence, internal fixation to maintain correction, and a solid spinal fusion of the entire length of the deformity to the sacrum.

The best age for surgery is debatable. Sharrard's (1968)¹¹ operation on neonates was unsuccessful because the cartilaginous

vertebrae at the osteotomy site failed to fuse and the deformity recurred. We now know that a long spinal fusion is necessary to stabilize the spine,³ but if this is performed at an early age it will cause extreme stunting of the trunk as the child grows to maturity. However, if kyphectomy and spine fusion are postponed to the teenage years, the deformity will have become too severe and



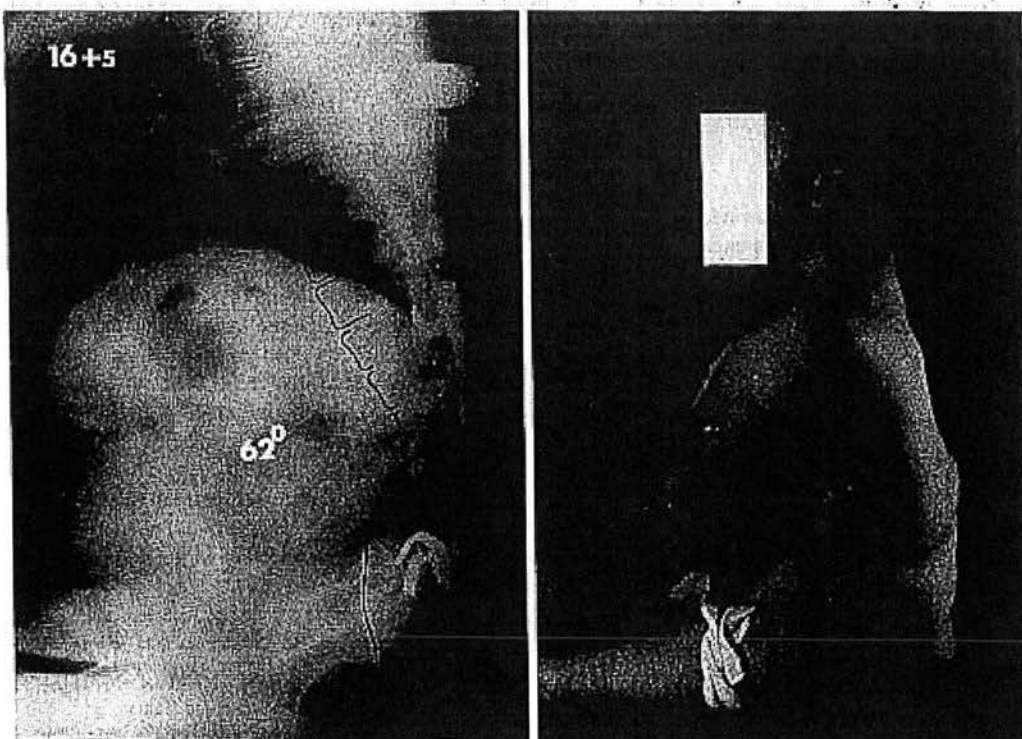
Figs 11,12. (Case 3) **11** (left), At 17 years 3 months, the kyphosis is 33° and good correction has been maintained. A mild thoracolumbar lordosis has formed above the fused lumbar area. **12** (right), The patient has a flat back without pressure sores and is able to sit upright without using his arms for support.



Figs 13-15. (Case 7) **13** (left), A boy aged 8 years 8 months with a severe lumbar kyphosis. **14** (middle), The kyphosis measures 132°. **15** (right), Following kyphectomy and internal fixation with two Harrington distraction rods from T9 to the sacrum and a single Dwyer cable, the kyphosis was reduced to 50°.

correction will be impossible. The best compromise is therefore to delay surgery until 8 to 10 years of age when the vertebrae are sufficiently developed to allow the application of effective internal fixation and a more successful fusion. Resection of the kyphosis is only indicated in very young children if there is difficulty in obtaining skin closure over the myelomeningocele, with the understanding that a second osteotomy is likely to be required at a later stage.

The kyphosis is extremely rigid and a flat back cannot be achieved without excising two or three vertebrae from the apex of the deformity. This is best performed through a posterior approach and necessitates partial resection of the nonfunctioning spinal cord at the level of the osteotomy. Such surgery is not without its risks and in this series one patient died during the procedure. In the nine surviving patients, the mean kyphosis before the operation was 131° (range 125° to 166°) and this was reduced by a mean of 66%



Figs 16, 17. (Case 7) **16** (left), At 16 years 5 months one of the Harrington rods has broken but there is a solid fusion and satisfactory correction has been maintained at 62°. **17** (right), The patient has a flat back without pressure sores and is able to sit upright without the support of his arms.

to a mean of 44° (range 4° to 70°), following the surgery. A flattening of the superficial soft tissues overlying the spine was achieved in all of these patients although it was rarely possible to achieve complete radiographic correction.

Although kyphectomy significantly reduces the bony prominence, it is difficult to maintain correction and also prevent the tendency for a thoracic lordosis to develop above the level of the osteotomy. Correction is best maintained by applying rigid internal fixation, and the most successful method in this series (Group 3 patients) was by means of a combination of three-point fixation using rigid Harrington rods applied posteriorly with pressure over the osteotomy site, restraining forces in the thoracic region and on the sacrum, and compression across the osteotomy site by means of staples or a single Dwyer cable (Figures 15,16). In more recent patients, the Harrington rods have been further reinforced by segmental sublaminar wiring in the thoracic region and wiring around the pedicles and through the vertebrae in the lumbar region where the laminae are deficient.⁴ A posteriorly applied AO plate also provided excellent correction and stabilization (Figures 10,11) but this necessitated a very wide resection of the dural sac and caused profuse bleeding from the epidural veins, which was difficult to control. Dwyer instrumentation when used alone was the most unsuccessful method of internal fixation (Cases 1 and 2) because it was not sufficiently rigid to maintain correction (Figures 5,6).

The long-term stability of the spine depends on producing a solid posterior spinal fusion extending from the mid-thoracic region to the sacrum. The three patients in Groups 1 and 2 had a spinal fusion confined to the lumbar region, and this did not prevent a thoracic lordosis developing above the fused area; in one of these patients (Case 1) the upper body fell forward once more, necessitating the use of the arms for support. In order to prevent this from happening, all six patients in Group 3 had a spinal fusion with instrumentation extending from the mid-thoracic region to the sacrum and all had a stable spine at skeletal maturity after a minimum follow-up of 7 years. All of these patients had a flat back without pressure sores and all were able to sit erect without using their arms for support (Case 7, Figures 13-17). In three of these patients, the degree of correction at final follow up was 7° to 18° better than it was immediately after surgery. In these patients there was a relatively localized anterior fusion confined to the region of the osteotomy, and it is possible that the improved correction was due to the continued anterior growth of the unfused vertebral bodies in the presence of the tethering effect produced by a long posterior fusion.

There is no ideal treatment for these unfortunate children. Despite having a stable spine, all of the children who were operated upon before the age of 10 years tended to obesity and had stunting of their trunks at skeletal maturity. However, this was no more than it would have been if left untreated. I therefore feel that although

kyphectomy is a very major procedure with a high incidence of complications, it has a definite place in the treatment of the myelomeningocele child with a kyphosis.

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THE LUMBAR THECO-PERITONEAL SHUNT SYNDROME AND ITS SURGICAL MANAGEMENT

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Seven patients treated in infancy by a lumbar theco-peritoneal shunt for idiopathic communicating hydrocephalus presented later in childhood after developing a characteristic orthopaedic syndrome. This included a severe, rigid and progressive lumbar hyperlordosis, severe bilateral restriction of straight leg raising and abnormalities of stance and gait.

Four of the patients, who had severe hyperlordotic curves of over 90°, required operations to correct their extreme deformity. The recommended method of correction is a three-stage procedure: first, anterior wedge resection osteotomies at several levels in the lumbar spine, then a period of "90-90" femoral traction, and finally a posterior fusion and stabilisation using Harrington instrumentation. The results were good, with few complications.

This paper describes the presentation and treatment of an orthopaedic syndrome which can develop many years after the insertion of a theco-peritoneal shunt for the treatment of idiopathic hydrocephalus in otherwise normal children. This shunt procedure drains cerebrospinal fluid from the lumbar subarachnoid space to the peritoneal cavity by means of a small polyethylene tube (Matson 1949). More recently, because of recurrent blockages, this shunt has been largely superseded by the more successful ventriculo-atrial shunt. However, some children who had theco-peritoneal shunts inserted many years previously have presented to the Edinburgh Scoliosis Clinic having developed a severe spinal deformity. These children were found to have a characteristic syndrome of extreme lumbar hyperlordosis (Fig. 1), bilateral limitation of straight leg raising and an abnormality of stance and gait. The incidence of this syndrome is not known. In the only reported long-term review, Kushner *et al.* (1971) examined 28 children several years after the theco-peritoneal shunting procedure and found that 13 had developed a scoliosis and that six of these had a lumbar hyperlordosis.

CLINICAL MATERIAL

Between 1973 and 1979, seven patients treated in infancy by lumbar theco-peritoneal shunt for idiopathic communicating hydrocephalus have presented at the Edinburgh Scoliosis Clinic with a severe lumbar hyperlordosis.

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There were four girls and three boys, aged between 8 and 16 years (mean 11 years) when they presented.

Theco-peritoneal shunt had been carried out on them between the ages of two months and 18 months (mean seven months). During the four years after the shunt, all the patients had required revision operations to renew the tube, which had become blocked, from once to four times in each patient (on average twice). In three patients the lumbar theco-peritoneal shunt was eventually abandoned and replaced by a Pudenz ventriculo-atrial shunt.

Symptoms. The earliest onset of symptoms was at a mean age of nine years, the most common initial complaint being increasing difficulty in walking. Three patients stated that they were able to walk only on their toes with their knees bent, having previously walked normally. The other four patients complained that they had a shuffling gait and could walk less far than formerly. Four patients complained of an increasing hollowing of their lower backs. Low backache was present in four patients and two had used a spinal support without improvement.
Clinical signs. All but one of the patients were of normal intelligence and attended normal schools. All seven had two major clinical abnormalities: an increased lumbar lordosis, and severe bilateral restriction of straight leg raising. Six patients also had some scoliosis but increased lumbar lordosis was the major deformity.

Lumbar hyperlordosis. The increase in lumbar lordosis was mild in two children, moderate in one and very severe in four. Clinical details of these severe cases are given in Table I, while photographs and radiographs of two of them are shown in Figures 1 to 12. The patients with severe hyperlordosis stood with marked protuberance of the abdomen and shortening of the trunk (Figs 1 and 7). The pelvis was rotated forward to such a degree that the anus faced posteriorly. The sacrum was hori-



Fig. 1

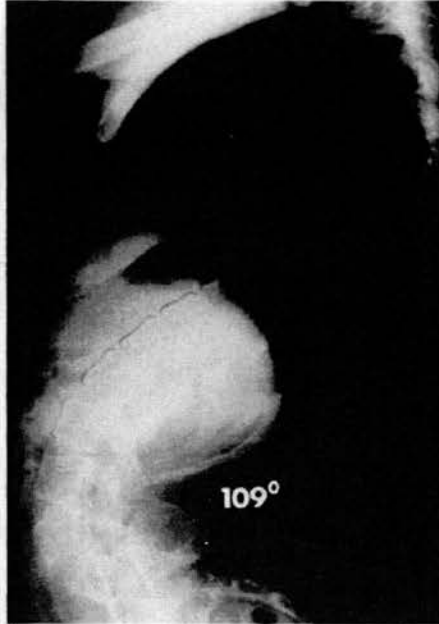


Fig. 2

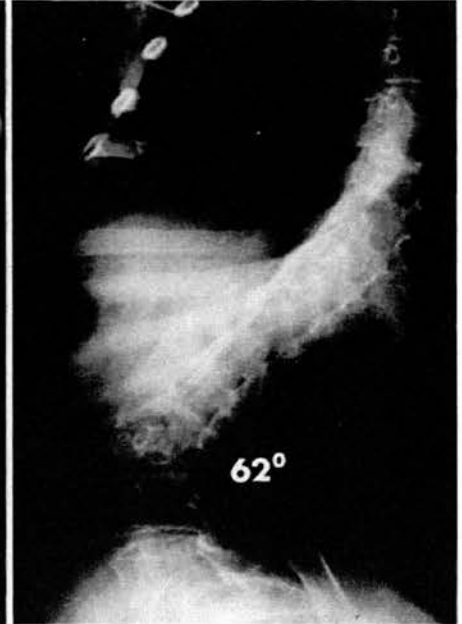


Fig. 3

Case 2. Photographs and radiographs before treatment. Figure 1—A girl aged 10 years with severe lumbar hyperlordosis. She can stand only on tiptoe with hips and knees flexed, because of the tightness of her hamstrings. Figure 2—A lateral radiograph, taken standing, shows a 109° lumbar lordosis with a horizontal sacrum (inclination 90°). Figure 3—A radiograph, taken standing, shows a 62° left lumbar scoliosis, causing the upper trunk to list to the right.



Fig. 4

After treatment. Figure 4—One year after anterior wedge osteotomies and fusion, the lumbar lordosis and posture are normal. The patient has gained 7.6 cm in height. Figure 5—At the age of 14 years, there is solid anterior spinal fusion from L3 to the sacrum. The lumbar lordosis and sacral inclination have remained normal. Figure 6—A radiograph at 14 years shows that the lumbar scoliosis which was only 14°, after her operations, is now 34°.

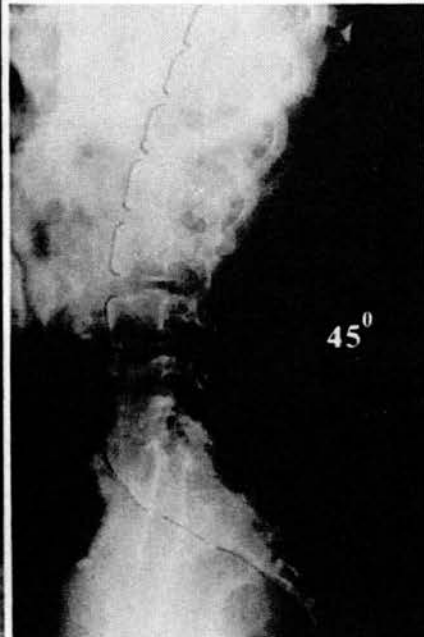


Fig. 5

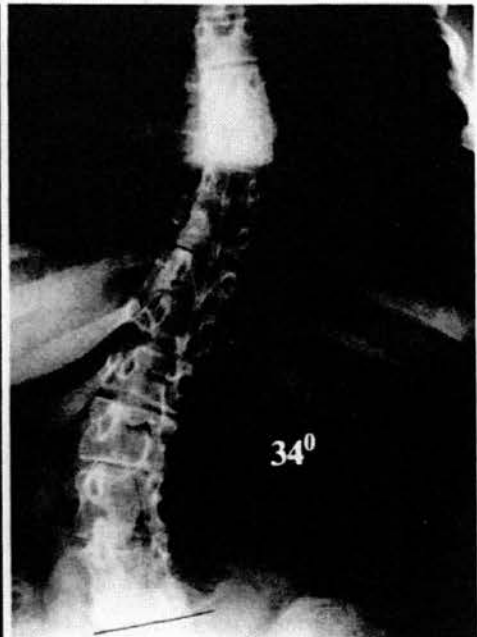


Fig. 6

Table I. Details of the four patients with severe lumbar hyperlordosis who had operative treatment

Case number	Before treatment		Sex	Lordosis	Sacral inclination	Scoliosis	Treatment
	Age						
	years	months					
1	16	3	M	T11 to S1 165	120	Right lumbar T7 to L5 52 List to right	Pre-operative halo traction Anterior lumbar osteotomies L2/3, L3/4 and L4/5
2 (Figs 1 to 3)	10	0	F	L1 to S1 109	90	Left lumbar T10 to S1 62 List to right	Anterior lumbar osteotomies L3/4, L4/5 and L5/S1 90-90 femoral traction
3 (Figs 7 to 9)	13	7	F	T11 to S1 150	98	Right thoracic T5 to T12 Slight list to right	Anterior lumbar osteotomies L3/4 and L4/5 90-90 femoral traction Posterior fusion with Harrington instrumentation L1 to S1
4	12	9	M	T11 to S1 92	88	Left thoracic T2 to T12 56 List to left	Anterior lumbar osteotomies L3/4, L4/5 and L5/S1 90-90 femoral traction

zontal and the apex of the lumbar lordosis was so deep that the posterior elements of the spine could be palpated only with difficulty. In compensation for the lumbar hyperlordosis all patients had a smooth thoracic kyphosis. The hyperlordosis was completely rigid and neither suspension nor forward bending produced any correction. The thoracic kyphosis was not as rigid and could be partly corrected by suspension. In five patients there was a tendency for the upper trunk to list to one side.

Scoliosis. Four patients also had a mild to moderate left lumbar scoliosis and two patients had a moderate right thoracic scoliosis. In four patients the upper trunk listed towards the concave side of the scoliosis. The list was most pronounced in those patients with a lumbar

scoliosis and became more severe when they attempted to bend forward at the hips.

Straight leg raising. All seven patients had severe limitation of straight leg raising due to tightness of their hamstrings. In the four patients with severe lumbar lordosis (over 90°) straight leg raising was zero, and in the other three patients it was 40° or less. Because of the tightness of their hamstrings, two patients were able to stand only on their toes with hips and knees bent (Fig. 1), and two other patients had to walk in this manner. In these four patients ankle dorsiflexion could only be achieved when the knee was flexed more than 90°. All the patients had a waddling gait, swinging the pelvis forward on the same side as the leading leg.



Fig. 7

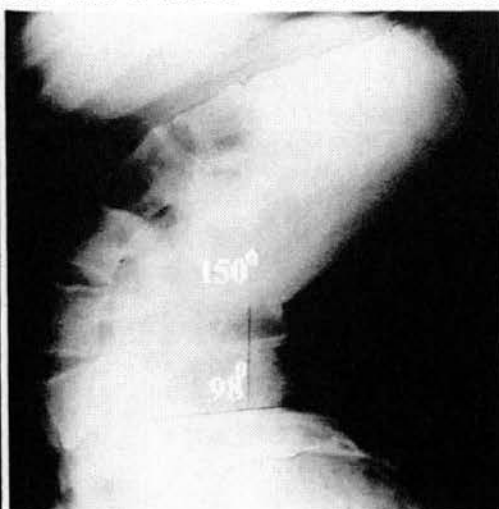


Fig. 8

Case 3. Photograph and radiographs before treatment. Figure 7—A girl aged 13 years and seven months with a very severe lumbar hyperlordosis. Figure 8—A lateral radiograph, taken standing, shows 150° of lumbar lordosis. The sacrum is angled forwards and downwards at an inclination of 98°. Figure 9—A radiograph, taken standing, shows a 46° right thoracic scoliosis and a slight list to the right.

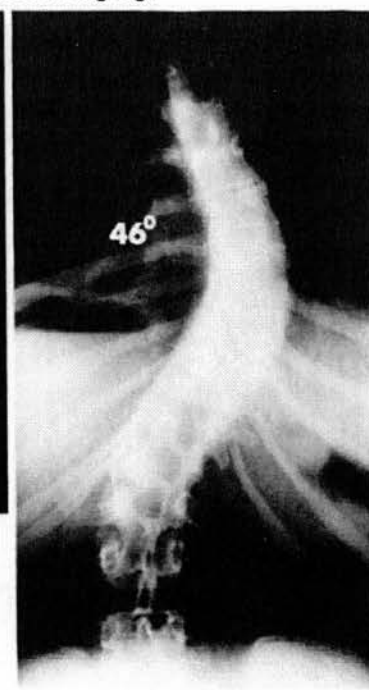


Fig. 9

Table II. The results of operative treatment of the four patients who had severe lumbar hyperlordosis

Case number	At the end of treatment				At final review				
	Lordosis	Sacral inclination	Scoliosis	Complications	Age		Lordosis	Sacral inclination	Scoliosis
					years	months			
1	T11 to S1 42°	56°	T7 to L5 30° No list	Permanent paralysis of left quadriceps since pre-operative traction	21	9	T11 to S1 55°	56°	T7 to L5 75° No list
2 (Figs 4 to 6)	L1 to S1 47°	47°	T10 to S1 14° No list	Bilateral weakness of ankle dorsiflexion and eversion and paraesthesia during traction after operation. Full recovery in six months	14	4	L1 to S1 50°	45°	T10 to S1 34° No list
3 (Figs 10 to 12)	T11 to S1 43°	48°	T5 to T12 39° No list	None	16	3	T11 to S1 40°	48°	T5 to T12 35° No list
4	T11 to S1 54°	48°	T2 to T12 32° No list	Thoracic scoliosis increased to 62° after one year. Posterior fusion with Harrington instrumentation from T4 to S1 carried out	16	2	T11 to S1 49°	40°	T2 to T12 30° No list

There were no motor or sensory disturbances and the only abnormal neurological finding was a moderate increase in the ankle and knee reflexes of one patient.

Radiographs. The angles of lordosis and scoliosis were measured, using the Cobb method (1948), on standard lateral and anteroposterior radiographs, taken with the patient standing. Lumbar hyperlordosis extended from the thoracolumbar junction to the sacrum and measured from 65° to 165° with a mean of 100°. Normal lumbar lordosis ranges from 40° to 60° (Moe *et al.* 1978).

Sacral inclination was measured on the lateral spinal radiograph using the method described by Wiltse and Winter (1983). Normal sacral inclination is approximately 45°; a horizontal sacrum would have 90° inclination. The inclination in our patients ranged from 72° to 120° with a mean of 85°. In three of the four patients with very severe lordosis, sacral inclination was over 90° and the upper sacral segments were angled downwards towards the floor (Fig. 8).

Lumbar scoliosis, present in four patients, extended



Fig. 10



Fig. 11

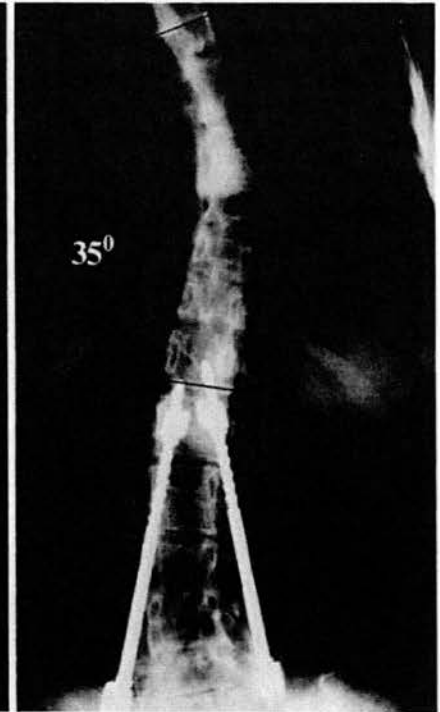


Fig. 12

Case 3. After treatment. Figure 10—One year after anterior and posterior correction and fusion, lumbar lordosis and sacral inclination are both normal. The patient has gained 15 cm in height. Figure 11—At the age of 16 years a lateral radiograph shows solid anterior and posterior fusion. Lumbar lordosis and sacral inclination have remained normal. Figure 12—The radiograph shows that the spontaneous correction of thoracic scoliosis has remained stable at 35°.

from the lower thoracic region to the sacrum (Fig. 3). The mean Cobb angle was 28° (range 20° to 62°). These curves were moderately rotated, and all were associated with a list towards the concave side. Two other patients had thoracic scoliotic curves (Fig. 9) of 46° and 65° with mild rotation.

Pelvic obliquity due to asymmetrical hip flexion was seen on the erect anteroposterior radiographs of two patients. This disappeared completely when the radiographs were repeated with the patients supine.

Myelography. Myelograms were performed on the four patients with severe hyperlordosis. Lumbar puncture was impossible because of the severity of the lordosis and Myodil had to be injected by cisternal puncture. In one patient a complete block was seen at the level of the tenth thoracic vertebra and was thought to be due to adhesions. In two there was a block at the apex of the lumbar lordosis which was partial in the supine position, but became complete when the patient was prone. Three patients showed loculation of the Myodil suggestive of arachnoiditis.

TREATMENT

The four patients with very severe hyperlordosis required operations in several stages to obtain complete correction of their extreme deformity. Three of these patients were operated upon by MJM.

Anterior spinal osteotomies. The lower lumbar spine was exposed anteriorly through a transperitoneal approach and the great vessels mobilised to either side. The lower three lumbar discs and parts of the adjacent vertebrae were excised as anteriorly based wedges which extended back to the posterior longitudinal ligament. Partial correction of the lordosis was obtained by flexion of the hips, with firm downward pressure on the spine.

Femoral traction. Complete closure of the anterior open wedges left by resection was obtained slowly over the first few days after operation, by applying increasing femoral traction with both hips and knees flexed to 90° (Fig. 13). If neurological abnormalities developed they could be detected early and the traction released.

Posterior spinal fusion and stabilisation. Posterior fusion and stabilisation of the spine by means of Harrington instrumentation was done about two weeks after the anterior operation. No further correction was attempted and the Harrington rods were used only as splints from the thoracolumbar junction to the ala of the sacrum on each side (Figs 11 and 12).

After this operation the patient was returned to an ordinary bed without traction. A well-fitting underarm lightweight jacket was applied when the wound had healed. The patient was then encouraged to walk and was allowed home when confident. The jacket was removed nine months later when the spine was solidly fused.

RESULTS

The details of the operative treatment and results of the four patients are shown in Tables I and II. Follow-up

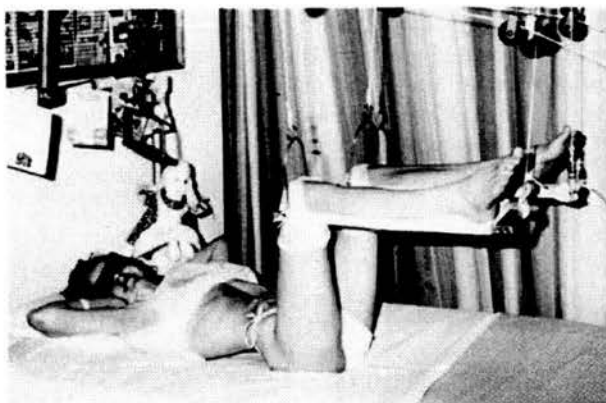


Fig. 13

After the first operation the patient is placed in "90-90" traction to close the open wedges of the anterior osteotomies.

was for a mean of four years (range 2 years 8 months to 5 years 6 months), by which time three patients had reached skeletal maturity.

Lumbar hyperlordosis. The mean lumbar hyperlordosis in the four patients before treatment was 129° (range 92° to 165°); soon after treatment this was reduced to 46° (range 42° to 54°). At final follow-up good correction had been maintained (Figs 5 and 11). The mean lordosis was 45°, and the range of 42° to 54° is within normal limits.

Sacral inclination. The mean sacral inclination at presentation was 99° (range 88° to 120°); after treatment this had been corrected to a mean of 50° (range 47° to 56°). At final follow-up there had been slight gain in correction and the mean sacral inclination was 47° (range 40° to 56°) which is within the normal range.

Scoliosis. Two patients (Cases 1 and 2) also had a lumbar scoliosis which was associated with a marked list of the upper trunk towards the concave side of the scoliosis. Both patients were treated by anterior wedge resection osteotomy and "90-90" traction. Neither had a posterior spinal fusion. The lumbar scoliosis and the list of the trunk were corrected significantly after these procedures, but during follow-up the scoliosis tended to recur (Table II).

A thoracic scoliosis was present in the other two patients with severe hyperlordosis (Cases 3 and 4). These curves corrected spontaneously after anterior osteotomies of the lumbar spine; because of this it was decided that neither curve required treatment. During follow-up one of these thoracic curves remained stable (Case 3, Fig. 12), but the other (Case 4), increased from 32° to 62° and after one year required correction by means of Harrington instrumentation and a posterior spinal fusion. The thoracic scoliosis was corrected to 30° and then remained stable.

Stance and gait. Before treatment all four patients tended to walk on tiptoe with knees flexed and a rolling gait. One patient could stand only with hips and knees flexed and heels off the ground (Case 2, Fig. 1). After correction of the lumbar hyperlordosis all four patients could stand

normally (Case 2, Fig. 4 and Case 3, Fig. 10), and walk with a heel-and-toe gait. However, straight leg raising was minimally improved and as a result they all still tended to walk with a rolling gait, swinging the pelvis forward on the same side as the leading leg.

The correction of the lumbar hyperlordosis in all the patients produced a marked gain in height, ranging from 5 to 15.3 cm. Three patients who had pain before treatment gained complete relief of their low backache after spinal surgery.

Complications. There were no deaths. Complete unilateral paralysis of the quadriceps occurred in one patient (Case 1), who had been treated with halo traction before operation. Traction was immediately released but the nerve palsy did not recover. After this experience, traction was not applied to any patient before operation. This was the only permanent and severe complication in the series.

Another patient (Case 2), developed bilateral weakness of dorsiflexion and eversion of the ankles with paraesthesia in the feet while in "90-90" traction after anterior spinal osteotomy. Traction was immediately reduced and no further correction was attempted. Second-stage posterior fusion was not performed for fear of increasing the neurological deficit. A complete neurological recovery took place over the next three months.

DISCUSSION

Infants who have had a lumbar theco-peritoneal shunt inserted for communicating hydrocephalus may present later in childhood having developed a characteristic syndrome which is not seen in any other condition.

The main deformity in all our patients was a severe lumbar hyperlordosis. This was first noted at a mean age of nine years and was progressive, giving an extremely ugly appearance. Normal lumbar lordosis ranges from 40 to 60 (Moe *et al.* 1978); in our severely affected patients the mean lumbar lordosis was 100° and the sacrum was rotated into a horizontal position. The cause of this very rigid deformity is unknown. It is possible that the insertion of the shunt, and the many revisions which were often necessary, could cause irritative arachnoiditis and tethering of the cauda equina to the bony structures. Then, as the child grows, a reflex protective spasm may develop in the posterior spinal muscles to produce an increasing lumbar lordosis which later becomes fixed. Of the other possible causes of hyperlordosis we found no evidence of interlaminar fusion or of significant scars in the paraspinal muscles.

The second component of the syndrome was very severe bilateral limitation of straight leg raising caused by tightness of the hamstrings. This prevented the child from striding forward normally by flexion of the hip and extension of the knee. The patients compensated for this restriction by swinging the ipsilateral side of the pelvis forward with the leading leg and limiting extension of the knee by walking on tiptoe. In our opinion this tightness

of the hamstrings was due to protective spasm resulting from arachnoiditis and limitation of movement of the cauda equina. After surgical correction of the hyperlordosis, both stance and gait were improved because of the relative lengthening of the hamstrings which occurred when the pelvis was rotated back into a more normal position.

The few published papers on the surgical correction of hyperlordosis in the theco-peritoneal shunt syndrome have reported a high incidence of serious neurological complications (Hall and King 1971; Steel and Adams 1972; Moe *et al.* 1978). In our series the anterior closing wedge osteotomies shortened the vertebral column, and so minimised the risk of stretching the tethered cauda equina. Complete correction was obtained postoperatively by slowly increasing femoral traction with both hips and knees in 90° of flexion. This technique allows neurological abnormalities to be detected early and reversed by releasing the traction. Pre-operative traction or first-stage correction by means of Harrington distraction rods are absolutely contra-indicated because of the very high incidence of serious neurological complications. We found that the anterior fusion stabilised the corrected hyperlordosis, but failed to prevent recurrence of lumbar scoliosis. To control the lumbar scoliosis a posterior spinal fusion was needed as a second stage. Harrington distraction rods were then used to hold the correction obtained by the anterior operation and post-operative traction, but no further correction was attempted for fear of stretching the tethered cauda equina. Thoracic scoliosis, when present, usually corrected spontaneously after the anterior operation and, because of this, the thoracic curve was not included in the posterior fusion. Any progressive thoracic scoliosis could be treated separately at a later date.

The methods reported here make possible the safe and satisfactory correction of the very severe spinal deformity which can occur as a result of a theco-peritoneal shunting procedure. In our opinion this neurosurgical procedure is now contra-indicated because of these serious orthopaedic complications.

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Surgical Correction of Spinal Deformities Following Spinal Cord Injury Occurring in Childhood

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This article reports on the surgical treatment of 14 consecutive patients with paralytic spinal deformities secondary to spinal cord injury occurring in childhood. Eleven patients underwent a posterior spinal fusion and three patients underwent a combined anterior and posterior spinal arthrodesis. Luque rods were used in all but one patient. The spinal fusion extended to the sacrum in 10 patients. No patient developed postoperative wound infections or medical complications. Four patients (28.6%) who underwent initially a posterior spinal arthrodesis developed pseudarthrosis. This was treated successfully by a combined anterior and posterior spinal fusion in two patients. The remaining patients underwent a revision posterior spinal fusion with recurrence of the nonunion in one patient. A combined anterior and posterior spinal arthrodesis could be considered the treatment of choice for patients with severe deformities who can tolerate anterior surgery. If pseudarthrosis develops following posterior spinal fusion, this can be best treated by a combined anterior and posterior revision procedure with instrumentation. (Journal of Surgical Orthopaedic Advances 16(4):174-186, 2007)

Key words: paralytic, scoliosis, spinal cord injury, spinal deformity, surgical treatment

Paralysis occurring in childhood is a devastating problem, which has both medical and psychosocial implications. A total number of 10,000 spinal cord injuries (SCIs) occur in the United States every year, which represents almost 40 patients per 1 million people (1-3). The mean recorded age at the time of neurological injury is around 30 years, with a percentage of only 3% to 5% of SCIs presenting in children younger than 15 years of age (4-6).

There is an increased mortality following a SCI, which has been reported to range up to 50% of the affected individuals (6). Children who have sustained SCIs have an overall mortality rate of 5% to 10% during the first

year after the traumatic incident, with the majority of these early deaths occurring as the consequence of cardiac or respiratory complications developing in the first 2 weeks following injury (7). Fatalities due to cervical spine injuries in children are directly correlated with the level of the lesion and the age of the patient. The more cephalad the cervical injury and the younger the child, the higher is the risk of early death (8).

After the acute phase of treatment, children who have complete or incomplete spinal cord deficits may develop a number of complications that can manifest in the short and long term and that are directly related to the paralysis. Spinal deformity affecting both the coronal and the sagittal balance of the spine and hip subluxation or dislocation are the most common orthopaedic problems in younger patients (9-11). Other causes of paralysis and subsequent spinal deformity in children include the development or the treatment of an intraspinal lesion, such as a neoplasm or an infection, and ischemic complications occurring during cardiothoracic or vascular surgery (12, 13).

The spinal deformity in the paralyzed child may result from a number of contributing etiological factors. These include a generalized trunk muscle weakness with or without associated spasticity, which causes the vertebral column to collapse, due to the effect of gravity, into a scoliosis, kyphosis, lordosis, or a combination of these deformities. This involves a large segment of the spine with the apex located usually in the thoracolumbar region and

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is commonly associated with a significant pelvic obliquity (14). Spinal deformity can also occur as the result of a direct injury to the vertebral growth plates, producing an asymmetric growth arrest. Vertebral column trauma can lead to the development of a chronic instability and a progressive deformity. Patients who undergo neurosurgical decompression or excision of an intraspinal lesion through multilevel laminectomies are prone to developing a postlaminectomy deformity, especially at or above the thoracolumbar junction (15, 16). Older children with a preexisting idiopathic scoliosis or kyphosis may experience significant curve progression when paralysis is superimposed (5).

The purpose of this study was to report on the surgical treatment of patients who developed a paralytic spinal deformity secondary to SCIs occurring in childhood. We believe that it is important to increase awareness of the complexity of these deformities in regard to their specific surgical considerations and the incidence of postoperative complications that occur in this particular patient population and can jeopardize an inherently challenging surgical procedure.

Materials and Methods

We reviewed the medical records and spine radiographs of 18 consecutive patients with a paralytic spinal deformity as a consequence of a SCI who were seen at our institution between 1980 and 2005. All patients were treated and followed by the senior author (MJM). There were 11 male and seven female patients. Fifteen patients had a scoliosis, one patient had a kyphosis and two had a lordoscoliosis. Patients who had underlying neuromuscular disorders, patients who underwent multiple-level laminectomies and developed a postlaminectomy deformity, and patients with a history of vertebral column fractures at one or more levels without spinal cord deficits were excluded from this study.

All 18 patients had developed deformities of the spine and were referred to our tertiary spinal deformity center for further treatment. Because we did not have access to the whole population of patients who sustained SCIs in childhood in our region, we are unable to draw conclusions on the prevalence or the severity of the spinal deformity that developed in this group of patients.

Preoperative Assessment

All patients underwent a thorough neurological examination at their first visit in the outpatient clinic to determine the level of their spinal cord paralysis. The site of the curvature was defined according to the classification

proposed by the Scoliosis Research Society (17): cervicothoracic (apex C7 or T1), thoracic (apex between T2 and T11), thoracolumbar (apex at T12 or L1), lumbar (apex between L2 and L4), and lumbosacral (apex at L5 or caudad). All of the serial spine radiographs for each patient in this study were measured by the senior author in both the coronal and sagittal planes with the use of the Cobb method (18) with the aim of eliminating inter-observer measurement error. Care was taken to measure all curves with use of the same anatomical landmarks on the serial spine radiographs. All spine radiographs were made with the patient sitting. Preoperative anteroposterior traction radiographs were obtained in every patient to assess flexibility of the spinal deformity and the associated pelvic obliquity. The flexibility of the deformity was also assessed with vertical suspension of the patients during the clinical examination.

Surgical Indications

Patients with a relatively flexible scoliosis or sagittal deformity, which would correct to at least 50° on an anteroposterior traction radiograph and without fixed pelvic obliquity, underwent a single-stage posterior spinal fusion with the Luque technique of segmental sublaminar fixation and the use of allograft bone. The spinal fusion extended to include the lumbosacral articulation in all patients with marked associated pelvic obliquity using the Galveston technique of intramedullary rod fixation in the iliac bones. Patients with rigid spinal deformities and increased comorbidities, primarily due to a poor cardiorespiratory function, would not tolerate a combined anterior and posterior spinal arthrodesis and they also underwent a single-stage posterior spinal fusion. A combined one- or two-stage anterior and posterior spinal arthrodesis was performed in those patients with severe and stiff spinal deformities who did not have significant preoperative cardiac or pulmonary compromise with the use of the Luque-Galveston technique of posterior segmental fixation with or without supplementary anterior stabilization of the spine.

Postoperative Care

The postoperative care consisted of a detailed clinical and radiographic review, which was performed at 3 weeks, 3 months, 6 months, and 1 year following surgery, and then on a yearly basis. The postoperative follow-up extended routinely until skeletal maturity for all patients treated surgically in childhood and adolescence, apart from those who were operated on later, and all patients underwent a minimum 2-year clinical and radiographic evaluation.

TABLE 1 Description of SCI, subsequent spinal deformity, and surgical treatment applied in this patient population

Patient No./ Gender	Etiology of SCI	Neurological Deficit	Age at SCI (yrs)	Type of Spinal Deformity	Age at Surgery	Surgical Treatment	Preoperative Curve Size	Postoperative Curve Size	Complications and Reoperations
1/M	Leukemia	Paraparesis T6	3	T kyphosis	11.6	PSF with Luque rods, T3-L5	110°	60°	1st nonunion: removal of Luque rods and regrafting, 2nd nonunion: PSF with AO USS instrumentation
2/M	Spinal trauma	Paraparesis T1	3	TL scoliosis	12.5	PSF with Luque-Galveston technique, T3-sacrum	86°	34°	Nonunion: combined single-stage A/PSF with posterior AO USS instrumentation
3/M	Spinal trauma	Paraparesis T3	2	TL scoliosis	15.8	PSF with AO USS II instrumentation and sublamina wires, T4-L5	73°	42°	Nonunion: combined single-stage A/PSF with anterior AO USS instrumentation
4/F	Spinal trauma	Quadriparesis	14	TL scoliosis	17	PSF with Luque-Galveston technique, T4-sacrum	73°	34°	Nonunion: PSF with AO USS instrumentation
5/M	Vascular injury during cardiac surgery	Paraparesis T6	4.2	TL lordoscoliosis	11.1	PSF with Luque-Galveston technique, T3-sacrum	lordosis: 147°, scoliosis: 56°	lordosis: 60°, scoliosis: 20°	
6/F	Spinal trauma	Paraparesis T3	9.8	TL scoliosis	17.6	PSF with Luque-Galveston technique, T3-sacrum	67°	30°	
7/M	Thoracic neuroblastoma	Paraparesis T4	0.1	TL lordoscoliosis	6.3	Combined 2-stage A/PSF with Luque-Galveston technique, T3-sacrum	lordosis: 110°, scoliosis: 81°	lordosis: 50°, scoliosis: 34°	
8/M	Thoracic neuroblastoma	Paraparesis T5	3	TL scoliosis	11.8	Combined single stage A/PSF with Luque-Galveston technique, T2-sacrum	127°	47°	
9/F	Spinal trauma	Paraparesis T2	4.8	TL scoliosis	8	PSF with Luque-Galveston technique, T4-sacrum	38°	23°	
10/F	Cervical astrocytoma	Quadriparesis	5	T scoliosis	11.8	PSF with Luque rods, T3-L4	105°	60°	
11/M	Cervical astrocytoma	Quadriparesis	4.3	T scoliosis	13.1	PSF with Luque rods, C7-L5	91°	29°	
12/F	Birth injury to the cervical spine	Paraparesis C7	birth	TL scoliosis	22.5	Combined 2-stage A/PSF with Dwyer instrumentation and Luque-Galveston technique, T5-sacrum	105°	50°	
13/M	Spinal trauma	Paraparesis T4	13.9	TL scoliosis	17.7	PSF with Luque-Galveston technique, T4-sacrum	58°	24°	
14/F	Meningitis and hydrocephalus	Paraparesis T1	9.5	T scoliosis	11.2	PSF with Luque-Galveston technique, T1-sacrum	81°	30°	

M, male; F, female; SCI, spinal cord injury; T, thoracic; TL, thoracolumbar; PSF, posterior spinal fusion; ASF, anterior spinal fusion; A/PSF, anterior and posterior spinal fusion.

Results

The etiology and type of SCI, the age at onset of neurological deficit, the type of spinal deformity, the age and type of surgical treatment, and the treatment outcome are shown in Table 1.

Etiology of SCI

The cause of paralysis in this group of patients included a traumatic incident in 10 patients, spinal cord tumor in six patients, vascular injury to the neural cord following cardiac arrest during heart surgery in one patient, and meningitis with the development of hydrocephalus and subsequent cerebral and spinal cord lesion in one patient. Three of the six patients with an intraspinal tumorous lesion had an astrocytoma, two had a neuroblastoma, and one patient had leukemia.

Neurological Deficit

Twelve patients presented with high- or midthoracic paraparesis, which was complete in all but two patients. Six patients developed tetraparesis, which was incomplete in three of these patients. None of our patients had any ambulatory function at the time of presentation and they were all wheelchair dependent. Four patients with tetraparesis and one patient with paraparesis had increased spasticity, which was asymmetric in one of the patients with tetraparesis.

Age at SCI

The SCI that created the permanent neurological deficit occurred before the age of 10 years (mean, 4; range, birth to 9.5 years) in 16 patients. There were two patients who sustained a traumatic incident affecting their vertebral column and resulting in a SCI at 14 years of age and still developed a long C-shaped thoracolumbar scoliosis necessitating surgical treatment.

Scoliosis

Fifteen patients developed an isolated long C-shaped neuromuscular scoliosis affecting a large segment of the spine with the apex located in the thoracolumbar junction in 12 of these patients. Eleven of these 12 patients had a significant associated pelvic obliquity of more than 10°. In three patients, the apex of the scoliosis was in the thoracic spine and they still developed a long neuromuscular curve without a compensatory deformity in the lumbar spine.

One of these three patients with the thoracic scoliosis had an associated pelvic obliquity of more than 10°.

Eleven patients with a scoliosis underwent surgical treatment with a mean preoperative Cobb angle of 82° (range, 38°–127°). Seven of these patients had a pelvic obliquity of more than 10° with a mean preoperative angle of 20° (range, 12°–35°).

Kyphosis

One patient presented with an increased long thoracic kyphosis measuring 110° at the time of the spinal fusion without associated coronal spinal imbalance or pelvic obliquity.

Lordoscoliosis

One patient developed an increased lumbar lordosis measuring 147° and a long thoracolumbar scoliosis measuring 56° at the time of the spinal arthrodesis. This was associated with marked fixed flexion deformity of the hips and an exaggerated anterior pelvic tilt measuring 30°. The patient had an incomplete paraplegia at the T6 level after cardiac surgery. He was our only patient who could initially walk short distances using bilateral callipers (therapeutic walker); however, over time he lost his ability to ambulate and became restricted to his wheelchair.

Another patient with paraplegia due to an intraspinal neuroblastoma presented with a thoracolumbar hyperlordosis measuring 110° and a thoracolumbar scoliosis spanning a long segment of the spine and measuring 81° with associated anterior pelvic tilt and a 60° flexion contracture of the hips at the time of spinal correction.

Surgical Treatment

Fourteen patients underwent surgical correction of their spinal deformities. None of these 14 patients had received conservative treatment in the form of an underarm thoracolumbosacral orthosis (TLSO). Four patients were referred elsewhere for surgical management of their scoliosis because of family relocation.

The mean age at spinal arthrodesis was 13.4 years (range, 6.3–22.5). The mean length of postoperative follow-up was 6.4 years (range, 2.2–17). Ten patients underwent operative treatment before the end of their spinal growth. All these 10 patients were kept under regular clinical reviews for more than 2 years postsurgery until they reached skeletal maturity. Surgical correction of the scoliosis was also performed in four patients whose spinal growth had been completed. These four patients had been followed up for more than 2 years after their spinal fusion. Patients who required repeat surgery to treat

a pseudarthrosis have been followed for more than 2 years after their latest operation to exclude recurrence of the nonunion.

Surgical Technique

Eleven patients underwent a posterior spinal arthrodesis alone and three patients underwent a combined anterior and posterior spinal fusion. This was performed as a single-stage procedure in one patient and in two stages in the remaining two patients.

Posterior spinal instrumentation with bilateral Luque rods and segmental fixation with sublaminar wires was used in all but one patient to correct the deformity and stabilize the spine. The posterior instrumentation extended proximally from the cervicothoracic junction to the upper thoracic spine, depending on the level of the apex of the spinal deformity. Distal fixation of the Luque rods to the pelvis using the Galveston technique of intrailiac stabilization was required in 10 patients. From the remaining four patients, three patients with a thoracic scoliosis or kyphosis did not have pelvic imbalance and one patient with a thoracolumbar scoliosis had an associated pelvic obliquity of less than 10°. Therefore, in these four patients the posterior instrumented spinal fusion did not include the lumbosacral junction and ended at L4 or L5 in order to preserve distal spinal mobility. In the patient with the thoracolumbar scoliosis whose spinal arthrodesis spared the lumbosacral joint (patient 3), this was performed with the use of the AO USS II posterior spinal instrumentation (AO-Universal Spine System, Stratec Medical, Oberdorf) and segmental fixation with pedicle screws distally and sublaminar wires proximally.

An anterior spinal release preceded the posterior instrumented spinal fusion in two patients with a thoracolumbar scoliosis and in one patient with a thoracolumbar lordoscoliosis who had very severe and rigid deformities. This was performed through a thoracoabdominal retroperitoneal approach to the spine and involved a complete discectomy and annulectomy, as well as the release of the anterior longitudinal ligament at multiple levels across the convexity of the scoliosis. Supplementary anterior fixation with the Dwyer instrumentation system was used in one patient with a thoracolumbar scoliosis who underwent a staged anteroposterior spinal arthrodesis (patient 12). A combination of locally harvested autologous and abundant allograft bone was applied in all patients to reinforce the spinal fixation and achieve a solid bony fusion across the instrumented levels.

Postoperative immobilization in a spinal jacket or a brace was not used for any of our patients following the initial surgical procedure. All patients received aggressive respiratory physical therapy following surgery and were mobilized in the immediate postoperative period on their

wheelchairs, which were adapted to accommodate for their corrected spinal posture.

Complications and Reoperations

None of the patients developed postoperative wound infections, either early or late. There were no major medical complications following spinal fusion in this group of patients that would result in a prolonged intensive care unit stay and increased hospitalization time.

Four of the 14 patients (28.6%) who had initially undergone a posterior spinal arthrodesis alone developed an asymptomatic pseudarthrosis with failure of the instrumentation (breakage of the Luque rods). The first patient (1/M, Table 1) had a thoracic kyphosis measuring preoperatively 110°, which was corrected to 60°, and developed a pseudarthrosis 2 years following a posterior spinal fusion. The broken Luque rods were removed and the nonunion was repaired with the use of allograft bone without instrumentation. The patient mobilized following the application of a spinal jacket for a period of 4 months after the second procedure. A persistent pseudarthrosis was diagnosed 2 years later as a result of further deformity progression. This was treated with regrafting of the site of the nonunion and posterior stabilization using AO USS II instrumentation. The patient was fitted with an underarm spinal jacket for a period of 4 months postsurgery. Follow-up at 2.5 years showed the kyphosis to measure 72° and no evidence of recurrent pseudarthrosis.

The second patient (2/M, Table 1) had a thoracolumbar scoliosis measuring preoperatively 86°, which was corrected to 34°, and developed a pseudarthrosis 1 year following a posterior spinal fusion (Fig. 1). The broken Luque rods were removed and a combined single-stage anterior and posterior spinal arthrodesis with repair of the nonunion was performed using AO USS II anterior and posterior instrumentation with a combination of autologous rib and allograft bone. The patient mobilized following the application of a spinal jacket for a period of 4 months after the second procedure. Follow-up at 5 years showed the scoliosis to measure 67° and no evidence of recurrent pseudarthrosis.

The third patient (3/M, Table 1) had a thoracolumbar scoliosis measuring preoperatively 73°, which was corrected to 42°, and developed a pseudarthrosis 2 years following a posterior spinal fusion (Fig. 2). A combined single-stage anterior and posterior spinal arthrodesis with repair of the nonunion was performed using AO USS II anterior instrumentation, autologous rib, and allograft bone. The broken rods were reattached with the use of end-to-end tubular connectors at the level of the pseudarthrosis. The sublaminar wires were removed at the site of the nonunion. The patient was fitted with an underarm

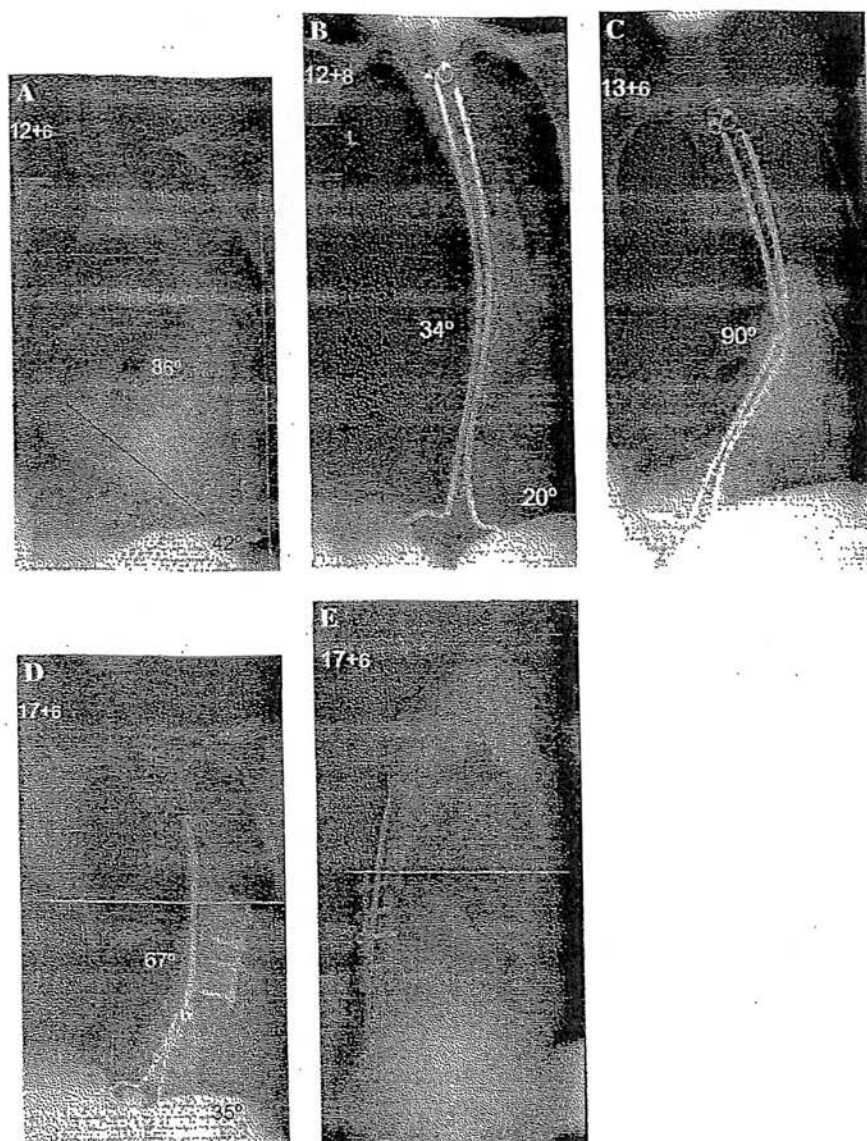


FIGURE 1 Preoperative anteroposterior radiograph of the spine (A) of a boy aged 12.5 years (patient 2) with a right thoracolumbar scoliosis measuring 86°, with 42° of associated pelvic obliquity (black line). The patient underwent a posterior spinal arthrodesis extending from T3 to the sacrum, which resulted in a satisfactory correction of his scoliosis to 34° and marked reduction of the pelvic obliquity to 20° (B). One year later he developed a pseudarthrosis at the apex of the curve with breakage of the Luque rods and recurrence of the deformity (C). This was treated with a combined one-stage anterior and posterior spinal fusion using AO USS II anterior and posterior instrumentation with a successful outcome in regard to healing of the nonunion but significant residual spinal and pelvic deformity measuring 67° and 35°, respectively (D, E).

spinal jacket for a period of 4 months postsurgery. Follow-up at 3 years showed the scoliosis to measure 67° and no evidence of recurrent pseudarthrosis.

The fourth patient (4/F, Table 1) had a thoracolumbar scoliosis measuring preoperatively 73°, which was corrected to 34°, and developed a pseudarthrosis 2 years following a posterior spinal fusion. A revision posterior spinal arthrodesis with repair of the nonunion was performed using AO USS II instrumentation and allograft bone. The broken Luque rods were removed along

with the sublaminar wires. The patient was fitted with an underarm spinal jacket for a period of 4 months postsurgery. Follow-up at 3 years showed the scoliosis to measure 60° and no evidence of recurrent pseudarthrosis. Three of the four patients (patients 2–4, Table 1) who were treated for a pseudarthrosis and had a spinal jacket applied developed pressure sores at the apex of the rib deformity adjacent to the convexity of the scoliosis. These pressures ulcers were managed conservatively and healed without further problems.

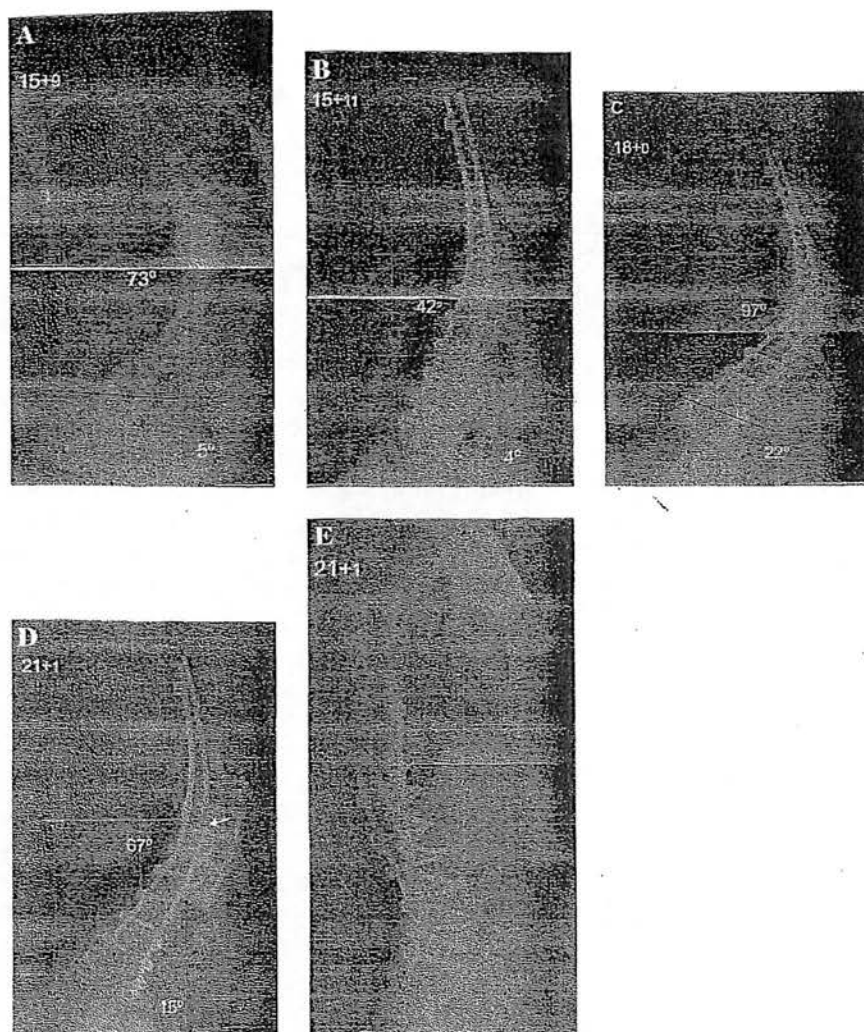


FIGURE 2 Preoperative anteroposterior radiograph of the spine (A) of a boy aged 15.8 years (patient 3) with a right thoracolumbar scoliosis measuring 73°, with 5° of associated pelvic obliquity (black line). Because of the minimum preoperative pelvic obliquity, the patient underwent a posterior spinal arthrodesis extending from T4 to L5, which resulted in a satisfactory correction of his scoliosis to 42° with 4° of residual pelvic obliquity (B). Two years later, he developed a pseudarthrosis at the apex of the curve with breakage of the rods, recurrence of the scoliosis, and add-on pelvic deformity measuring 22° (black line) (C). This was treated with a combined one-stage anterior and posterior spinal fusion using AO USS II anterior instrumentation with a successful outcome in regard to healing of the nonunion but a significant residual spinal and pelvic deformity measuring 67° and 15°, respectively (D, E). Note that the end-to-end tubular-connector that was used to reattach the posterior rods was disconnected on the right side without adverse effects on the progress of the bony fusion (white arrow).

Surgical Outcome

In the 11 patients with a scoliosis, the curve was corrected by 55.5% to a mean of 36.6° (range, 23°–60°). This correction was maintained at follow-up in all patients apart from the three patients who developed a pseudarthrosis. In the seven patients with a significant pelvic obliquity, this was corrected by 60% to a mean of 8° (range, 5°–15°). The correction of the pelvic imbalance was also maintained at follow-up.

In the two patients with a lordoscoliosis, the lordosis was corrected by 61%, from a preoperative mean of 128.5° to a postoperative mean of 50°. The associated

scoliosis in these two patients was corrected by 60.5%, from a preoperative mean of 68.5° to a postoperative mean of 27°.

Discussion

The development of a spinal deformity, usually affecting the coronal and occasionally the sagittal balance of the spine is a recognized complication of paralysis following a SCI occurring in childhood. The most significant factor that influences the incidence and severity of paralytic scoliosis is the age of the patient at the time

of the onset of paralysis, with younger patients being at a significantly higher risk (5). If a child sustains a SCI more than a year before reaching skeletal maturity, there is a 98% chance of developing scoliosis and a 67% chance of necessitating surgical treatment (19–21). In contrast, a SCI that occurs less than a year before skeletal maturity carries only a 20% risk of developing scoliosis, with an associated 5% chance of progressing to surgery (19). Mayfield et al. (20) reported that all their patients who had a SCI before the adolescent growth spurt presented with spinal deformities, which were most commonly causing a scoliosis, with 68% of these patients requiring surgical treatment.

Lancourt et al. (21) reported an incidence of scoliosis of up to 100% in children affected by a SCI before the age of 10 years, with 80% of these patients developing a severe curve, which the authors defined as greater than 40°. In the same study, only 19% of children who were paralyzed between 10 and 16 years of age, and 12% of children older than 17 years developed a severe scoliosis (21). Dearolf et al. (19) recorded an incidence of scoliosis of 97% in children injured before their adolescent growth spurt, and 52% in those patients who were paralyzed after their rapid phase of growth. The segmental level of the SCI does not appear to be related to the risk of developing a spinal deformity in patients with complete or incomplete neurological deficits (19). Lancourt et al. (21) also found no direct correlation between the neurological level of injury and the incidence of scoliosis; 86% of their patients with a cervical and 78% of those with a lower thoracic SCI developed a scoliosis.

In our series, the most common cause of paralysis was related to a traumatic SCI (10 of 18 patients; 55.5%). The causes of SCIs in children vary primarily with the age of the child and the type of activities. The most frequent causes of a SCI in the young population include falls and motor vehicle accidents (22–24). Falls even from a low height (less than 5 feet) can cause a significant vertebral column injury, especially affecting the cervical spine in the younger children (less than 6 years of age) (25–27). An injury to the cervical spine is more likely to produce severe permanent neurological complications and subsequently carries a higher risk of developing a posttraumatic spinal deformity compared with injuries involving more distal segments of the vertebral column.

Motor vehicle accidents affecting children most commonly involve passengers of automobiles, pedestrians, or bicyclists run down by vehicles. The use of car seatbelts and airbags constitute major advances in reducing mortality from automobile-related accidents. However, if these are used improperly, they may actually increase the incidence of a SCI in children (28–30). High-velocity injuries resulting in open fractures of the vertebral column carry a worse prognosis than low-velocity

injuries causing closed spinal fractures. Sports-related injuries — including wrestling, football, horseback riding, and diving accidents — account for a smaller percentage of SCIs in children and adolescents (4, 31–33). Occasionally, a traumatic injury to the spine can occur during birth or can be the manifestation of shaken baby syndrome, and this can result in a significant neurological deficit and the later development of a scoliosis (34).

The second largest group of patients presenting with paralytic spinal deformity in our series were those with an underlying spinal cord tumor (6 of 18 patients; 33.3%). Tumorous lesions affecting the intraspinal structures can be classified according to their location within the vertebral canal into intramedullary (within the substance of the cord, such as an astrocytoma, ependymoma, or medulloblastoma) and extramedullary. The latter can be further subdivided into intradural (lying within the thecal sac, such as a neurofibroma, meningioma, or leukemia) or extradural (lying outwith the thecal sac but within the confines of the spinal canal, such as a neuroblastoma, Ewing's sarcoma, or leukemia).

The most common presentation of a spinal cord tumor includes a progressive weakness in the lower limbs that may lead to paralysis (12, 35, 36). Onset of paralysis occasionally occurs suddenly, due to an ischemic stroke affecting the spinal cord, which may be caused by extramedullary tumor compression, intraspinal hemorrhage, or bony collapse. Other less common clinical manifestations include the development of back pain, radicular pain, gait abnormalities, or a spinal deformity (37). Frequently, the clinical onset of signs and symptoms is subtle, and, therefore, the diagnosis of the neoplasm can be considerably delayed (38).

A spinal cord tumor that creates significant neurological compromise in childhood can lead to the development of a spinal deformity. In addition, the neurosurgical excision of these lesions also carries a significant risk of exacerbating preexisting neurological deficits caused by the neoplastic lesion or creating paralysis in a neurologically intact patient (39, 40). In a previous series of adult patients who underwent resection of spinal neurofibromas, the risk of an iatrogenic long-term neurological deficit was 7% affecting motor and 21% affecting sensory function (41). Patients who underwent multilevel laminectomies for removal of an intraspinal neoplasm are prone to developing a localized kyphosis (15, 16). This deformity is the consequence of marked posterior element instability and can occur regardless of any associated neurological deficit as the result of the tumor or the surgical treatment. Children who have received radiotherapy for a spinal cord tumor are also at higher risk of developing a postradiation spinal deformity. A high dose of radiation that exceeds 3000 rads, as well as an asymmetrical exposure of the

spine to the radiotherapy, have been shown to be significant risk factors (35).

In children and adolescents who present with a spinal deformity following a previous history of spinal neoplasm, the anticipated long-term life expectancy will determine whether surgical correction of the deformity should be considered. In these patients, a multidisciplinary preoperative approach, including primarily a detailed oncological assessment, cannot be overemphasized.

Development of a Paralytic Spinal Deformity

All children who have sustained SCIs resulting in paraparesis or tetraparesis should be assumed to be developing spinal deformities unless proved otherwise. Therefore, these patients need to be kept under regular clinical and radiographic observation to detect a pathological curvature of the spine at an early stage and apply appropriate treatment. The typical paralytic scoliosis involves a single, long, C-shaped curvature with the apex usually at the thoracolumbar junction associated with pelvic obliquity. Twelve of the 15 patients (80%) with a scoliosis in the present study developed a long thoracolumbar curvature, which was followed by significant pelvic imbalance in 11 patients (73.3%). None of our patients developed a scoliosis with a double curve pattern. This finding was similar to the series presented by Lancourt et al. (21) in which 25 of 31 patients (80.6%) with paralytic scoliosis had a single, long neuromuscular curve and no compensatory deformity.

Unlike idiopathic scoliosis, progression of the paralytic spinal deformity is rapid and may occur even before the adolescent growth spurt, especially in the presence of muscle spasticity (42). In addition, children with spasticity are at an increased risk of developing progressive acetabular dysplasia leading to hip subluxation or dislocation, either unilateral or bilateral. Factors that predispose to hip instability in this group of patients include marked spasticity, especially if this is asymmetric, preexisting pelvic obliquity, hip flexion and adduction contractures, or a flaccid paralysis which results in gradual displacement of the femoral heads out of the acetabulae. It is rare for these patients to have functional limitations or pain as a consequence of their hip deformity and to require surgical reconstruction of the hips. Soft tissue releases, operative reduction, and bony reconstruction can improve sitting, hygiene, and ease of care but are usually unnecessary (10). The development of an abnormality of the hips can, however, result in an exacerbation of the pelvic obliquity and the paralytic scoliosis. Conversely, progressive scoliosis may lead to pelvic imbalance and continue a vicious circle.

Functional Limitations

The development of a pathological spinal curvature in addition to the inherent neurological deficit as a consequence of the SCI may significantly restrict the patient's functional capacities and markedly increase the need for nursing care. Ambulatory patients may require walking aids and eventually become wheelchair-dependent as the spine deformity progresses. Wheelchair-bound patients gradually lose their sitting balance and become hand-dependent sitters. As their sitting tolerance is increasingly diminished, these patients have to rely on their arm and hand support to maintain an upright position. Trunk decompensation may ultimately lead to pain caused by impingement of the ribs against the iliac crest on the concavity of scoliosis. The development of pressure sores is not uncommon in children with skin insensitivity (21). Cardiac and pulmonary complications may also develop, especially if the curve size is so severe that the patient can hardly spend any significant amount of time in the erect or sitting position.

Prevention of the development of a neuropathic spinal deformity in children with paralysis caused by a SCI is not usually possible. At least half of the children who were injured in their preadolescent years will necessitate later spinal fusion (9, 21). However, anatomical reduction and stabilization of the initial vertebral column injury is always advisable to reduce the risk of a localized post-traumatic deformity. Measures to reduce spasticity and prevention of joint contractures in the lower extremities may also be helpful.

Conservative Management

Orthotic treatment in paralytic spinal deformity has been ineffective to halt curve progression (19). However, the application of an underarm brace in young children with an early paralytic scoliosis may be useful in slowing down curve progression and delaying surgical correction for a later stage, while at the same time allowing for further spinal growth (19, 21). A well-padded and perfectly fitted TLSO in this group of children may improve their sitting balance and preserve upper limb function (28). Care needs to be taken when using a spinal brace in patients with sensory deficits, because they are prone to developing pressure sores. Treatment should be initiated when the curve reaches 20°, even though there is no objective data to support this intervention. There is no role for bracing in children who are obese or who have rigid curves. In these patients, the application of the spinal orthosis will have no effect in the scoliosis and will increase significantly the risk of developing pressure ulcers of the skin. None of our patients was fitted with a TLSO because of the severity of their spinal deformity.

at initial presentation and they were all listed for surgical treatment. However, we would advocate the use of a well-molded bi-valved underarm brace as a temporary measure in young children with flexible curves and no fixed pelvic obliquity.

Surgical Management

Spinal arthrodesis with the use of segmental instrumentation is the only treatment modality that has a well-documented positive impact in children and adolescents with paralytic spinal deformities. Surgical treatment in this group of patients is indicated in the presence of curve progression and a curve size of 45° to 50°, ideally in children 10 years of age or older, especially if there is deterioration in their functional skills (42, 43). It is not always possible to delay surgery, particularly in patients with marked spasticity and a rapidly progressing curve in whom spinal fusion may be required at an earlier age.

Restoration of the coronal and sagittal spinal alignment and leveling of the pelvis should be considered equally if not more important aims of the surgery than the percentage of curve correction. The objectives of the surgical correction are to achieve trunk balance, improve respiratory function, alleviate the pain caused by impingement of the ribs against the iliac crest on the concave side of the curve, provide better sitting tolerance in wheelchair-bound patients, prevent the development of pressure sores, and retain standing and walking ability in ambulatory patients, thereby maximizing their overall level of function.

The spinal fusion should extend from the upper thoracic region, to prevent the development of a recurrent proximal kyphosis, to the pelvis, especially if pelvic obliquity is present, if the curve involves the sacrum, or if the patient has a poor sitting balance. Preserving distal spinal motion segments by fusing the spine short of the lumbosacral joint can theoretically maintain function in activities of daily living and transfers but may lead to caudal add-on deformity and exacerbation of preexisting pelvic imbalance. A shorter spinal arthrodesis that does not extend to the sacrum may be indicated, however, in those few patients with incomplete neurological lesions who are able to walk or in patients with minimal or no pelvic deformity. In our series, the spinal fusion extended to the sacrum with pelvic fixation in 10 of the 14 patients (71.4%). None of our patients had retained ambulatory function, but the remaining four patients in whom the spinal arthrodesis stopped at L4 or L5 did not have any significant preoperative pelvic obliquity. However, one of these four patients (3/M, Table 1) who was fused distally to L5, developed a pseudarthrosis which resulted in recurrence of his scoliosis and an add-on pelvic deformity (Fig. 2).

Anterior Spinal Surgery

In patients with fixed deformities, an additional anterior spinal release performed through multilevel discectomies is indicated to maximize the flexibility of the curvature and allow for increased correction when the posterior instrumentation is applied. A combined anterior release and posterior instrumented fusion can be performed under one or two anesthetic sessions and is indicated when curve correction to 50° to 60° cannot be obtained on suspension or traction radiographs or in the presence of a rigid pelvic obliquity (42). The combined anteroposterior procedure can achieve better curvature correction and decrease the incidence of pseudarthrosis and recurrent deformity through a circumferential spinal fusion. This was required in three of our patients because of severe and stiff spinal deformities.

Anterior surgery is also indicated if there is posterior element deficiency as a consequence of extensive laminectomies and to prevent the crankshaft phenomenon in children younger than 10 years of age. There was only one patient in our series who was treated with an isolated posterior spinal fusion at the age of 8 years, and there was no evidence of a crankshaft effect with the patient being followed up to skeletal maturity. In the presence of a significant neuropathic kyphosis, an anterior fusion is usually necessary in addition to the posterior procedure and can increase the chances for a successful arthrodesis.

Spinal Instrumentation

The development of the concept of segmental spinal fixation with the application of translational corrective forces through the use of multilevel sublaminar wires and two single rods by Luque (44) in 1977 revolutionized the surgical treatment of neuromuscular scoliosis. This technique achieved a wide distribution of forces over each vertebra, with an increased initial spinal stabilization and a low risk of instrumentation failure. The Galveston technique of intramedullary placement of the rods in the iliac bed was developed by Allen and Ferguson (45) and accomplished a secure pelvic fixation (46). All patients in the present study were treated with the Luque posterior segmental instrumentation system and the Galveston technique of intrailiac pelvic fixation.

More recently, newer instrumentation systems using primarily pedicle screws have been introduced in the management of neuromuscular and paralytic scoliosis. These are based on the same principle of segmental fixation with pedicle screws or hooks and the alternative of iliac bolts for lumbopelvic or sacroiliac plates for sacropelvic fixation. A common problem in the application of third-generation instrumentation that uses pedicle hooks or screws in patients with paralytic spinal deformities

who lack the ability to walk and, therefore, have marked associated osteopenia is poor vertebral fixation, which can limit the ability to perform corrective maneuvers and may increase significantly the risk of pseudarthrosis.

It has been our experience that in this group of patients with poor bone mineral quality, the laminae provide the strongest points of fixation compared with the pedicle or the vertebral body and can withstand segmental translational forces applied through sublaminar wires with the aim to achieve correction of the deformity and balance the spine in both the coronal and the sagittal planes. In contrast, if repeat surgery is required (e.g., to address a non-union), it is technically considerably easier and safer to revise an instrumentation system that uses pedicle screws with or without hooks as opposed to sublaminar wires. If the Luque instrumentation was used at the original surgery and the patient needs a revision because of the development of a pseudarthrosis, we would advocate the addition of an anterior spinal fusion with instrumentation to the posterior repair of the nonunion. Overall, we believe that there are no published data to support any possible benefits obtained by the use of third-generation instrumentation systems in the surgical treatment of paralytic spinal deformities compared with the traditionally used Luque-Galveston technique.

Surgical Outcome

The mean preoperative scoliosis angle in the current study was 82°, and this was corrected to a mean 36.6° following spinal fusion. Lancourt et al. (21) reported on 10 patients with a mean scoliosis of 71°, which was corrected to 31° after spinal arthrodesis. Mayfield et al. (20) presented a series of 23 patients with a mean scoliosis angle of 56°; 17 of these patients (74%) underwent surgical treatment due to progression of their deformity but the average correction was not reported.

Patients with SCI and a paralytic spinal deformity are often nutritionally and metabolically compromised and may have poor muscle tone and associated spasticity. They may also have poor vasomotor tone resulting in increased intraoperative bleeding during scoliosis correction. The use of a cell-saver can reduce the need for homologous blood transfusion during spinal fusion. Respiratory impairment and chronic recurrent urinary tract infections are other factors that can jeopardize the spinal procedure. These patients will have a greater risk of developing postoperative complications compared with children with an idiopathic scoliosis or other neuromuscular conditions. The complications include delayed wound healing and wound infection, atelectasis and consequent pulmonary infection, and pseudarthrosis (5, 21, 47).

Early mobilization of the patients on their modified wheelchairs and aggressive respiratory physical therapy

will reduce the risk of medical complications in the postoperative period. External orthotic support to the spine is usually not necessary and was only used in our study following the revision procedures to address a nonunion in four patients. However, three of these four patients who were treated for a pseudarthrosis and had a spinal jacket applied developed asymptomatic pressure sores adjacent to the apex of the rib prominence on the convex side of the scoliosis.

Complications

In our series of 14 patients with paralytic deformities who underwent spinal arthrodesis, there were no major postoperative medical complications. However, we recorded a high rate of pseudarthrosis (28.6%), with four patients requiring revision surgery after an initial posterior spinal fusion. The nonunion was treated successfully in two of these four patients with a combined anterior and posterior spinal fusion. The repair of the pseudarthrosis was performed through a repeat posterior spinal fusion in the remaining two patients and one of these patients necessitated a second revision procedure to address recurrence of the nonunion. In those three patients where a combined anterior and posterior spinal arthrodesis was performed at the time of the initial surgery, there was no evidence of a nonunion at the latest follow-up.

This increased rate of nonunion recorded in our series is comparable to previous reports. In the study presented by Mayfield et al. (20), 9 of 17 patients (53%) with paralytic scoliosis developed a pseudarthrosis following spinal fusion with Harrington rods. In five of these nine patients there were multiple sites of nonunion, which were most frequently located in the lumbar and lumbosacral spine (20). After these initial results, the authors have performed staged anterior and posterior spinal arthrodesis with the use of Dwyer or Zielke instrumentation anteriorly and Harrington rods posteriorly, which reduced the rate of pseudarthrosis to less than 20% (20).

In conclusion, children with SCIs and their families should be informed of the increased risk of developing a progressive spinal deformity, especially if the neurological injury has occurred before adolescence. These patients should be kept under close observation in order to detect the spinal decompensation at an early stage and apply appropriate treatment. Prophylactic bracing can improve sitting balance and allow for upper limb function. This should be instituted early in young children in the hope of slowing progression of the curvature and delaying surgical treatment for a later age. Spinal fusion is the only treatment that can correct the deformity, restore spinal balance, and maintain function. This is associated with an increased risk of perioperative complications and

multiple technical difficulties. Good preoperative planning and the use of a meticulous surgical technique, including segmental stabilization of the spine and the pelvis using the Luque-Galveston technique supplemented by abundant bone graft, have shown in our series to produce satisfactory results. In view of the high rate of nonunion recorded in the present study, we would recommend a combined anterior and posterior spinal arthrodesis as the initial treatment for patients who are at a good general medical condition to tolerate anterior surgery and who have severe deformities. If pseudarthrosis develops following an isolated posterior spinal fusion, this can be treated more effectively by a combined anterior and posterior revision procedure with the use of instrumentation, which can increase the chances for a successful outcome.

Summary

A retrospective study of 18 consecutive patients was performed to investigate the outcome of surgical correction in patients who developed a paralytic spinal deformity secondary to spinal cord injury occurring in childhood. A rate of nonunion of 28.6% was recorded in patients who underwent an isolated posterior spinal arthrodesis. This finding suggests that a combined anterior and posterior spinal arthrodesis could be considered the treatment of choice in patients who can tolerate anterior spinal surgery and who have severe deformities. If pseudarthrosis develops following a posterior spinal fusion alone, a combined anteroposterior revision procedure may be required to achieve a bony fusion.

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Neuromuscular Problems in Orthopaedics

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Chapter 7

The Management of Spinal Deformity in Neuromuscular Disease

M. J. McMASTER

Spinal deformity occurring as a consequence of neuromuscular disease can present one of the greatest challenges to the orthopaedic surgeon. These patients are very different from those with idiopathic or congenital scoliosis and must be assessed differently to avoid serious errors in management. In neuromuscular disease the treatment of the spine is influenced by a number of factors, such as respiratory muscle paralysis, sensory disturbance and functional disability of the limbs, which are not present in other types of scoliosis. Once a neuromuscular spinal deformity develops, it usually progresses and frequently requires treatment. If left untreated, the spinal deformity often becomes very severe and this may further decrease the respiratory function and make sitting and walking even more difficult.

Any neuromuscular disease producing weakness or spasticity of the trunk musculature can cause a spinal deformity provided the patient is still growing. In the past, poliomyelitis was the major cause of neuromuscular spinal deformity. Immunization has now virtually eradicated this disease from developed countries but there are many Third World countries where the condition remains a serious problem. Myelomeningocele and cerebral palsy are the commonest causes of neuromuscular spinal deformity in developed countries. In the past, many children with myelomeningocele rarely survived infancy, or early childhood, but more recently with early closure of the spinal defect, reliable means of controlling the hydrocephalus and antibiotics to control infection, an increasing number are surviving to adulthood when a spinal deformity

becomes their major problem (Hall & Bobechko 1973; Piggott 1980). These children along with those who develop a deformity following spinal cord injury are the most difficult to treat because of their sensory impairment. Other less frequent causes of spinal deformity are Friedreich's ataxia, spinal muscular atrophy, syringomyelia, and Duchenne muscular dystrophy.

In order to formulate a plan of treatment, it is necessary to understand the problems associated with the different patterns of neuromuscular spinal deformity and their course if left untreated.

Patterns of spinal deformity

In general there are two main groups of spinal deformities depending on the severity and distribution of muscle weakness affecting the spine. A third and much rarer type of spinal deformity occurs only in certain patients with myelomeningocele.

1 The commonest and most characteristic spinal deformity associated with neuromuscular disease develops in patients with either total paralysis or extensive symmetric weakness of their spinal musculature. Because of the lack of muscular support the spine 'collapses' under the influence of gravity when the patient is upright. Initially the spine remains flexible and the deformity can be easily corrected by passive means but with time it becomes fixed and increasingly more rigid.

The typical collapsing spinal deformity is a long smooth thoracolumbar scoliosis extending from the upper or mid thoracic regions to the

sacrum and producing pelvic obliquity. In addition to the scoliosis, the spine is also frequently deformed in the sagittal plane with an increased lumbar lordosis and occasionally a thoracic kyphosis (Kilfoyle *et al.* 1965). When sitting the combination of a severe scoliosis and pelvic obliquity frequently causes the patient to list to one side and necessitates the use of one hand for support. This interferes with the use of the arm for normal activities and further increases the degree of disability. The pelvic obliquity causes the patient to sit with most of the body weight over one buttock and this can result in a pressure sore especially if there is sensory impairment. When standing the fixed pelvic obliquity produces an apparent leg length discrepancy and this makes fitting of long leg calipers and walking even more difficult. Because of the pelvic obliquity the hip on the high side of the pelvis is not fully covered by the acetabulum; this predisposes to subluxation or dislocation especially if there is a flexion adduction deformity of the hip as a result of muscle imbalance.

2 The second group is neuromuscular spinal deformities due to asymmetric weakness of the spinal musculature. This imbalance results in a wide variety of curve patterns depending on the distribution of the muscle weakness and can affect any part of the spine from the cervical to the lumbar regions. These curves which can resemble idiopathic scoliosis may be either single or double and are not necessarily associated with pelvic obliquity. Initially the spinal deformity is flexible, but with time it becomes fixed and there may also be decompensation of the trunk.

3 An uncommon, but very characteristic, angular kyphotic deformity can occur in some patients with myelomeningocele and is present although not always recognized at birth. These patients have a total paraplegia and the defect in their laminae which extends throughout the lumbar region is much wider than in the majority of patients with myelomeningocele (Hoppenfeld 1967). As a result the paraspinal muscles which normally extend the spine are carried forward and act at a mechanical disadvantage (Drennan

1970). Once the child begins to sit, the kyphus which usually has its apex at the second or third lumbar vertebrae deteriorates rapidly under the influence of gravity and becomes very severe at an early age (Sharrard & Drennan 1972). This progression is only stopped mechanically when the ribs come to rest on the pelvic brim. The child is thrown forward and forced to use both hands for support. Sitting or lying is difficult and the anaesthetic scarred skin over the apex of the kyphus easily develops pressure sores.

Prognosis

Neuromuscular spinal deformities frequently develop in early childhood and all deteriorate with growth; this deterioration becomes most rapid during the pubertal growth spurt. After skeletal maturity the rate of deterioration slows but unlike idiopathic scoliosis the spine does not stabilize and frequently progression continues until it is stopped mechanically when the ribs abut on the pelvis or the patient becomes so disabled that he cannot sit upright.

Thoracic curves distort the rib cage and if severe will decrease the vital capacity and further interfere with the respiratory function which may already be affected by paralysis of the intercostal muscles. In thoracolumbar curves, the diaphragm may be the only functioning respiratory muscle, and this can be hindered by the upward pressure of the abdominal contents as the spine collapses. Patients may die from respiratory failure precipitated by their increasing spinal deformity and not due to progression of their neuromuscular disease.

Factors affecting treatment

Progressive deterioration of the neuromuscular disease

The majority of neuromuscular diseases are static although the spinal deformity may progress. In a few conditions such as Friedreich's ataxia

and Duchenne muscular dystrophy, the neuromuscular disorder may also progress resulting in increasing muscle weakness and finally death from cardiac or respiratory failure. Major surgical procedures on these patients necessitating bed rest and immobilization for more than a few days can result in a rapid increase in muscle weakness. As a result, patients who could walk prior to surgery, may no longer be able to do so afterwards. If spinal surgery is indicated it should be carried out at a relatively early stage before the patient is severely disabled and the period of bed rest should be as brief as possible.

Sensory impairment

In patients with myelomeningocele and spinal cord injury, the loss of sensation over their trunk and lower limbs will make them particularly liable to pressure sores. This is an important factor when deciding on the type of external support which may be necessary for conservative treatment or following surgery.

Respiratory function

The effect of a spinal deformity on respiratory function is very important when considering treatment. Many patients will already have an impaired respiratory function due to paralysis of their respiratory muscles and this cannot be altered by treatment. Further deterioration of the respiratory function can occur due to progression of the spinal deformity but this can be prevented by treatment. Care should be taken that any external support for the spine should not impair respiratory function by constricting the chest or limiting the movement of the diaphragm. Anterior spinal surgery may be hazardous because it involves a thoracotomy and taking down the diaphragm which may be the only functioning respiratory muscle. A very careful assessment of the respiratory function and blood gases is necessary prior to surgery. Unfortunately there is no single test which indicates a

safe level for surgery. Postoperative complications are less likely if the vital capacity is greater than 40% of the predicted normal and there is an effective cough to get rid of secretion. If the vital capacity is less than 30% spinal surgery will be hazardous and postoperative assisted ventilation will be necessary (Nickel & Perry 1961; Makley *et al.* 1968; Bonnett *et al.* 1975).

Cardiac function

Duchenne muscular dystrophy is associated with cardiac myopathy and this may also occur but not so frequently in Friedreich's ataxia (Hensing & MacEwen 1976). These patients require careful pre-operative cardiac assessment and monitoring during surgery.

Hip deformities

A unilateral contracture of the iliotibial band may contribute to pelvic obliquity (Irwin 1949) and this should be released prior to treating the spinal deformity. Hip flexion contractures will produce lumbar lordosis whereas hip extensor contractures predispose to hyperflexion of the pelvis and a lumbar kyphosis.

Mental function

Patients with cerebral palsy or myelomeningocele may also suffer from mental deficiency and this may make it difficult for them to understand or comply with a complex treatment regimen.

General condition

Many children with neuromuscular disease are unable to walk satisfactorily and as a result become obese. This makes it difficult to apply an external support to the spine and may also inter-

fere with surgery. Patients with myelomeningocele and spinal cord injury frequently have bladder paralysis and their renal function must be carefully assessed and any infection treated prior to surgery.

Radiographic evaluation

A careful radiographic evaluation as well as a clinical examination is necessary before treatment. Radiographs showing the whole of the spine in both the anteroposterior and lateral planes are taken with the patient standing and sitting. These radiographs show not only the severity of the scoliosis but also any kyphosis or lordosis that might be present as well as pelvic obliquity and decompensation of the trunk. The sitting radiograph eliminates the effect of crutches, hip flexion contractures or leg length discrepancy. A radiograph taken with the patient suspended or in traction will show the flexibility of the curves and this is important when deciding on the type of surgical procedure necessary to correct the spine.

Treatment

The key to successful management of neuromuscular spinal deformities is to recognize their poor prognosis at an early stage and to start immediate treatment while the curve is still small and before it becomes rigid. The only satisfactory conservative means of treatment is to brace the spine, but unfortunately this is never sufficient as a definitive treatment. Even if the deformity were successfully controlled to skeletal maturity, the curvature would again deteriorate once the brace was removed and progress to a severe degree. Consequently a spinal fusion is usually necessary at the end of brace treatment.

Posterior spinal fusion is an established and successful method of preventing increasing spinal deformity, but unfortunately cannot be satisfactorily applied to very young children. Neuromuscular curves are often very long and

the extensive fusion that is necessary will stop all longitudinal growth in the posterior elements resulting in a severe degree of stunting of the spine as the child grows to maturity. Bonnett *et al.* (1975) also found that an increasing lordosis could develop in young patients due to the continued growth of the vertebral bodies anteriorly and lack of growth in the fused posterior elements. In order to minimize these difficulties it is best to delay spinal fusion until just before the onset of puberty when the growth spurt makes the curves increasingly more difficult to control in a brace. Because neuromuscular curves often start at an early age, a long period of conservative treatment may be necessary to prevent deterioration before they can be satisfactorily fused at the optimum age between 10 and 12 years. However, an earlier fusion will be necessary if the deformity cannot be controlled by other means.

Brace treatment

The object of treating neuromuscular curves in a brace is to minimize their otherwise relentless rate of progression and to maintain good alignment and sitting balance until an adequate amount of spinal growth has occurred and there is adequate bone stock for a spinal fusion. Unlike the treatment of idiopathic scoliosis these patients with weakness of their trunk muscles cannot perform corrective spinal exercises while in their brace. The brace is used therefore only as a holding device with the object of producing a stiff spine in a corrected position. No attempt is made to mobilize the spine out of the brace and no exercises are performed. Although the brace may provide an initial partial correction this is usually not maintained and the curve continues to deteriorate but at a much slower rate than before the brace was applied.

Indications

The brace must be applied before a fixed spinal deformity develops. There is no value in apply-

ing the brace after the patient has developed a large structural curve and especially if there is pelvic obliquity (Duval-Beaupere *et al.* 1975). Any patient with a curve of over 25 degrees which shows signs of progression is a candidate for spinal bracing.

Contraindications

There are no absolute contraindications to bracing a neuromuscular scoliosis although there are a number of factors which mitigate against a successful result. Curves of over 60 degrees and especially those with pelvic obliquity are very difficult to control in a brace. The holding pressure of the brace against the skin is often uncomfortable and can result in pressure sores especially if the skin is anaesthetic. Extreme obesity and lack of maternal care makes brace therapy much more difficult and the likelihood of pressure sores much greater. There are also certain types of neuromuscular disease which do not respond well to brace therapy. Patients with cerebral palsy, Friedreich's ataxia, and Duchenne muscular dystrophy, may already have difficulty in walking and the application of a brace may totally immobilize them. If, however, these patients are already wheelchair bound, the application of a brace can be much more beneficial. In some patients, breathing may be difficult because of respiratory muscle paralysis and the constriction of a brace around their chest may further increase their difficulties by limiting chest expansion.

A brace should not be applied to myelomeningocele patients with an angular kyphotic deformity because this is totally unresponsive and the brace is a potent cause of pressure sores.

Types of spinal brace

1 The *Milwaukee brace* has been most frequently used in the conservative treatment of idiopathic scoliosis and has also been successfully applied to neuromuscular curves

(Bunch 1975). The Milwaukee brace consists of a carefully moulded plastic pelvic girdle connected to an encircling neck piece by extendable rods which provide a distracting force. A second correcting force is provided by means of an adjustable pad applied over the rib hump. Because these patients are paralysed, and cannot perform exercises while in the brace, the constant pressure of the pad may deform the ribs and also cause skin breakdown. In order to overcome these problems, the rib pad should be larger than normal and carefully conform to the chest wall. The pelvic girdle may be extended proximally to support the torso. The most frequent cause of failure of Milwaukee brace treatment is its application to curves which are too large. This type of brace is best applied to thoracic curves of less than 40 degrees and in patients with normal sensation.

2 An *underarm 'total contact' brace* (Fig. 7.1)

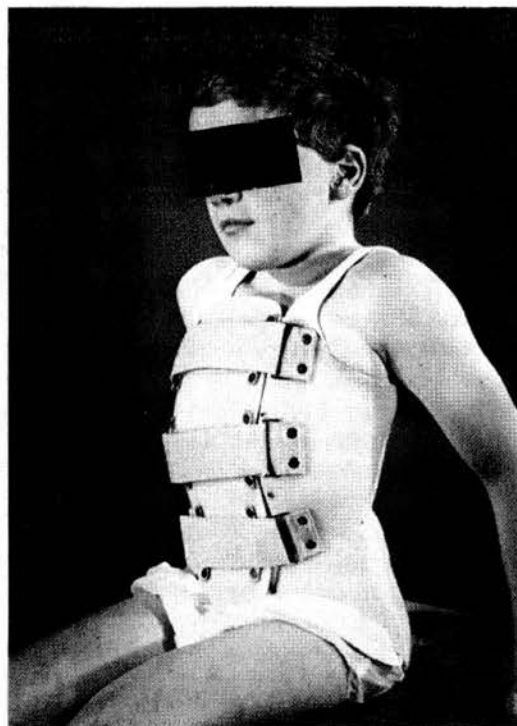


Fig. 7.1. A total contact underarm orthosis.

can be applied to the collapsing type of thoracolumbar scoliosis. This brace is not the same as that used in idiopathic scoliosis where a pressure pad within the brace provides part of a three point correcting system of forces. If this brace were applied to patients with reduced sensation it is likely to produce pressure sores. Patients with neuromuscular curves and reduced sensation are best treated in a total contact type of brace which spreads the pressure over a wide area so reducing the likelihood of skin breakdown. Care should be taken to make sure that these braces do not further compromise an already impaired respiratory system. Drennan *et al.* (1979) have produced a thoracic suspension orthosis which is a moulded body jacket suspended from a wheelchair by a swivel bracket. The thorax and subcostal margins act as weight-bearing structures and the effect of gravity on the suspended lower trunk corrects a collapsing scoliosis. The buttocks are kept just off the wheelchair and this prevents pressure sores.

Brace manufacture

Both the Milwaukee brace and the underarm jacket are made from a plaster mould of the patient held in the corrected position by applying traction to the head and pelvis. In Duchenne muscular dystrophy the underarm jacket is made in such a way as to extend the spine as this has been found to be the most stable position in preventing further deterioration (Wilkins & Gibson 1976). Patients with spinal muscular atrophy walk with a hyperlordotic thoracolumbar spine and the brace must be manufactured to maintain this position. If the brace were made with the lordosis corrected, as in idiopathic scoliosis, the patient would fall forward.

Both the Milwaukee brace and underarm jacket are made from strong but lightweight plastic materials and are lined with a nonabsorbent cushioning material to distribute the forces. The jacket is bivalved and secured with Velcro straps which allow easy removal for regular skin inspection.

Brace supervision

The patient is slowly weaned into the brace over a period of several weeks and this allows time for the skin to adapt to the pressure of the brace. The orthosis is worn only when upright and not at night when vertical support for a collapsing spine is unnecessary. If the patient were to wear the brace while asleep he is much more likely to develop pressure sores especially if there is anaesthetic skin. Good maternal care is essential and the brace should be removed several times each day to check for excessive pressure against the skin.

The satisfactory bracing of a neuromuscular spine deformity is difficult and requires meticulous attention. The best results are obtained when the spinal surgeon and the orthotist regularly see the patient together and the brace is made in close proximity to the clinic so that immediate alterations can be carried out as required.

Termination of brace treatment

Brace treatment may be terminated for several reasons. Firstly, if the curve cannot be controlled in the brace to an acceptable degree, it should not be allowed to progress and become fixed. Secondly, the brace may have to be removed if it produces intractable pressure sores. Thirdly, the patient may reach the optimum age for fusion, that is 10–12 years of age, and as fusion is always necessary one should not wait any longer. These curves are not like idiopathic scoliosis where if braced to skeletal maturity the spine becomes stable. Once the brace is removed from a neuromuscular scoliosis, the only option is to fuse the spine.

Operative treatment

Spinal fusion is frequently required to control neuromuscular spinal deformities. Before fusing the spine it is usual to correct the deformity as much as possible and then to hold the spine in the

corrected position until the fusion is solid. A successful spinal fusion transforms the abnormally curved part of the spine into a solid bar of bone that is of sufficient strength to resist bending under the influence of gravity and the deforming forces of muscle imbalance. Correction may be lost due either to the development of a pseudarthrosis or to bending of a solid but weak fusion.

In the last 25 years there have been many major advances in the treatment of spinal deformities and there is now virtually no severity of curvature which cannot be significantly improved and stabilized by surgery. However, it is important to balance what is technically possible and what is practicable and safe for these patients, many of whom are severely disabled by their neuromuscular disease. Pre-operative evaluation is vitally important and the timing and indications for surgery require much more consideration than simply the presence of a spinal deformity. The spine must not be treated in isolation and the other problems due to the neuromuscular disease must be taken into consideration.

Indications for surgery

The surgery required for these patients is often the most difficult type of spinal surgery and should only be carried out in a specialized centre by a surgeon experienced in dealing with all types of spinal deformity. Any patient over the age of 10 years with a progressive structural curve should be considered a candidate for spinal fusion. If surgery is thought necessary it should be carried out early to prevent problems from developing, and fusion should not be delayed until after the adolescent growth spurt.

In deciding on surgery, a loss of function is more important than the size of the curvature. A progressive pelvic obliquity which becomes fixed may lead to progressive decompensation of the trunk, a loss of sitting balance and pressure sores. If the patient is less than 10 years old and being treated in a brace, the curve should not be

allowed to exceed 60 degrees and should never go beyond 100 degrees. The pulmonary function may already be impaired by respiratory muscle paralysis and the addition of a severe spinal deformity will only increase this problem. A severe curvature may also require anterior surgery via a thoracotomy and this may not be possible if there is a severe respiratory deficiency. It is important therefore to carry out any spinal surgery before the pulmonary function becomes affected by the deformity. Older patients can develop backache due to their curvature and this may be a relative indication for surgery. The cosmetic appearance is not a primary indication for surgery, but it is much better for the patient's morale to be able to sit upright rather than be bent and twisted.

Contraindications

The main contraindication to spinal surgery is a severe impairment of respiratory function which makes it likely that the patient would develop postoperative respiratory failure and this would not recover with assisted ventilation. Other relative contraindications are a short life expectancy due to cardiac or renal problems and severe mental retardation which would significantly interfere with the postoperative care.

Objectives of surgery

The objective of surgery is to correct the deformity and fuse the spine in its optimum position. The trunk should be supported in a balanced position over a stable and level pelvis. The torso should be vertical with the head centred over the thorax and pelvis. This will allow the patient to sit in a stable position and have independent use of the arms for activities other than supporting the trunk. In this position the weight is symmetrically distributed over the buttocks and there is less likelihood of pressure sores. In patients with total paraplegia the ability to walk in long leg calipers is frequently improved fol-

lowing a long spinal fusion to the sacrum. In these patients, the normal functioning muscles in the upper body are better able to control the pelvis and lower limbs through the long stable lever of the fused spine. A 'swing-to' gait with crutches and long leg calipers may be improved to a 'swing-through' gait which is more economical. However, in some patients with only partial paralysis of the lower limbs and trunk, the long fusion to the sacrum may interfere with gait. These patients often walk in calipers by alternately hitching up each side of their pelvis to get the foot off the ground and swinging the leg through. A lumbosacral fusion and rigid spine will prevent tilting the pelvis and interfere with this gait. A trial in a plaster jacket before surgery may be useful in evaluating any alteration in gait pattern that might occur after spinal fusion.

Selection and extent of the fusion area

The most common error in the surgical treatment of neuromuscular spinal deformity is to fuse too short a segment of the spine. The extent of the fusion is determined by examining the erect sitting and standing spinal radiographs in both the anteroposterior and lateral planes. This enables one to assess the whole spinal deformity which may be any combination of a scoliosis, kyphosis and lordosis. The fusion should extend over the entire length of these deformities as seen on both the anteroposterior and lateral radiographs. The upper level of the fusion is often well above that chosen for idiopathic curves and is usually one or two vertebra above the end vertebra as measured on the radiographs. The top of the fusion should lie directly above the centre of the sacrum and all rotated vertebrae should be included in the fusion. If the upper level of the fusion is not high enough, the curve will continue to progress above the fused area. In a collapsing spine with pelvic obliquity it is always necessary to extend the fusion distally to the sacrum. In curves due to asymmetric paralysis and without pelvic obliquity the spine should be fused to the lowest neutral vertebra and it is not necessary to

extend the fusion to the sacrum. The extent of the fusion area for any curve does not change whether the patient is operated on anteriorly or posteriorly.

Pre-operative correction

Methods of correcting the spinal deformity may be applied either before surgery or at the time of fusion. It is not possible to apply pre-operative corrective plaster jackets safely to patients with neuromuscular spinal deformities because of the high risk of pressure sores developing beneath the plaster especially if the skin is anaesthetic. The more successful means of correction are applied intraoperatively at the time of spinal fusion. The intraoperative techniques of correction can be applied either to the posterior bony elements or anteriorly to the vertebral bodies.

Halofemoral and halopelvic traction

Pre-operative halofemoral or halopelvic distraction has been recommended for the correction of large fixed curves (Bonnett *et al.* 1972; O'Brien *et al.* 1975) but the degree of correction is usually no more than can be obtained by intraoperative means alone (Lonstein & Akbarnia 1983).

There are a number of disadvantages of pre-operative traction. While in traction the patient is immobilized in bed during which time the vertebrae may become more porotic and this makes the application of metallic internal fixation devices to the bone less secure. Halo-traction applied to a patient with a meningocele can be dangerous because the pins in the halo easily perforate the thin bone of a hydrocephalic skull. The presence of the pelvic pins passing through the wings of the ilium in pelvic hoop traction make it difficult to extend the fusion to the sacrum and also to obtain satisfactory bone grafting material from the iliac crest.

An advantage of halofemoral traction is that it can, on occasion, improve a limited vital capacity by reducing the intra-abdominal pressure so allowing the thoracic cavity to expand and making the patient a better operative risk. The traction may also be helpful in controlling an uncooperative patient and facilitating nursing care.

Posterior fusion and Harrington instrumentation

In the 1950s Harrington of Houston devised a technique of posterior spinal instrumentation which he first used to correct the scoliotic curves of patients with poliomyelitis (Harrington 1962). Since this time surgeons have used the technique in all types of neuromuscular spinal deformities with successful results.

Technique of Harrington instrumentation

Harrington instrumentation consists of two parts. A distraction system which jacks the curve out and supports the spine by means of a rigid stainless steel rod applied across the concavity and fixed to the spine by means of a hook at either end. These hooks can be applied to the spine anywhere from the first thoracic vertebra to the sacrum. A second and more flexible rod may be applied around the convexity and gains further correction by means of a system of hooks which apply compression to the spine. A transverse traction rod has also been devised to connect the distraction and compression rods and provides even greater stability. Harrington at first used his technique as a method of correcting the deformity without fusing the spine or applying external support. Unfortunately, the unrestricted movement caused the metallic hooks to cut out of the bone and it became obvious that a posterior spinal fusion with external immobilization was necessary if correction was to be maintained.

In neuromuscular collapsing curves, a long distraction rod is necessary and usually extends from the upper thoracic region to the ala of the sacrum on the high side of the pelvic obliquity. In severe curves a second distraction rod may also be applied in parallel nearer to the apex of the curve and this helps to distribute the distraction force and gains better correction (Fig. 7.2[a] and [b]). The distraction rod must also be prebent in the sagittal plane to correct and accommodate for a thoracic kyphosis and maintain a normal lumbar lordosis. Unfortunately, contouring the rod in the sagittal plane significantly decreases the amount of distraction force that can be applied. In order to prevent the contoured rod from rotating out of its sagittal alignment a square ended rod is used and inserted into a lower hook with a square hole. To correct the pelvic obliquity it is necessary to apply a downward force on the high side of the pelvis and a specially adapted lower hook has been designed to fit over the ala of the sacrum. Unfortunately, as the distracting force corrects the scoliosis it may also cause a flattening of the normal lumbar lordosis. This is especially disadvantageous in a patient with hip flexion contractures who can walk independently, because it throws the body forward and makes it difficult to stand and walk without support. A lumbar lordosis may be maintained by bending the distraction rod to conform to the lumbar spine and by applying a compression system to the convexity of the curve before applying distraction. Osteoporotic bone is more easily fractured, and as a result displacement of the hooks is more frequent than in idiopathic scoliosis. In order to prevent displacement of the upper distraction hook, its fixation site may be reinforced with methyl methacrylate cement.

Posterior fusion

The incidence of pseudarthrosis following posterior spinal fusion in neuromuscular spinal deformity is higher than in idiopathic scoliosis. Bonnett *et al.* (1975) in a study of the evolution

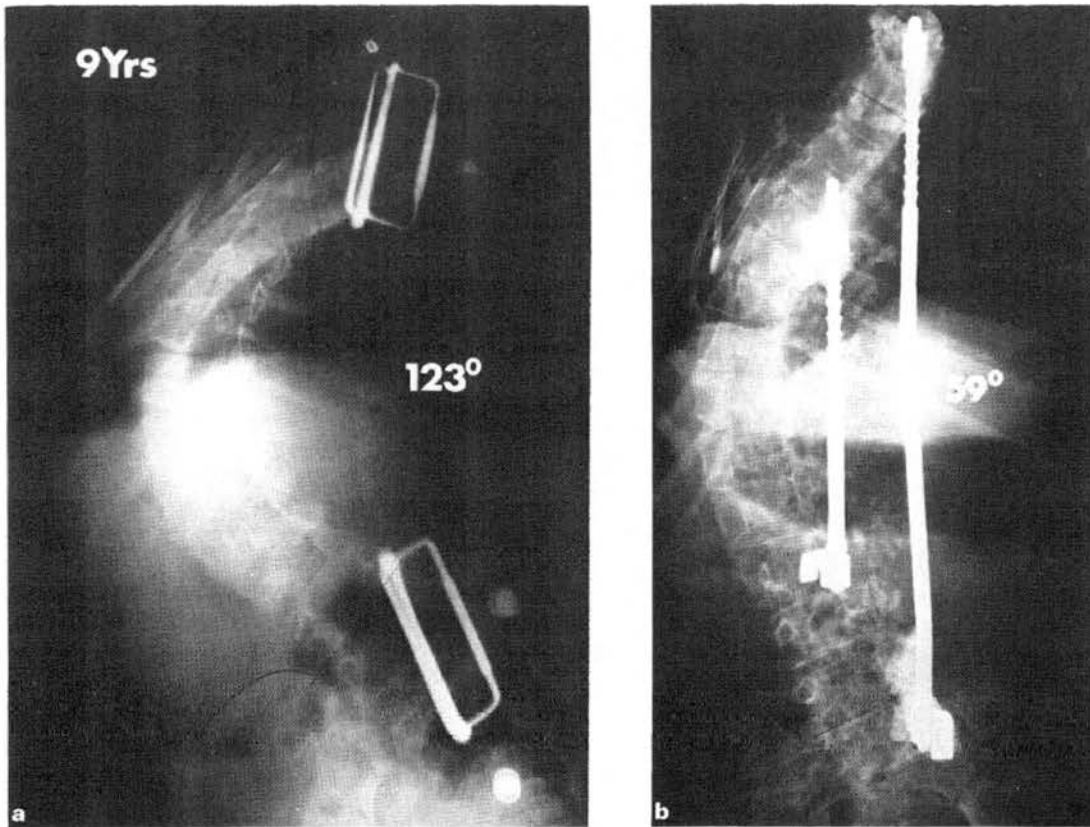


Fig. 7.2. (a) Anteroposterior erect spinal radiograph of a nine-year-old girl with spinal muscular atrophy. She has a collapsing thoracolumbar scoliosis measuring 123° and severe pelvic obliquity. (b) Using two Harrington distraction rods, the curve has been corrected to 59° and the pelvis levelled.

of their treatment of paralytic scoliosis at Rancho Los Amigos Hospital, found that despite Harrington instrumentation and a careful fusion technique, the incidence of pseudarthrosis was 27%. This compares with a less than 1% incidence of pseudarthrosis in patients with idiopathic scoliosis (Erwin *et al.* 1976). Special care, therefore, is required in fusing the spines of patients with neuromuscular deformities.

If the patient has a myelomeningocele scoliosis, the skin overlying the spine will be scarred and adherent to the dura. Great care is necessary in dissecting the skin from the dura and some surgeons prefer two parallel incisions over the facet joints and strip the tissues laterally

leaving intact the scarred and potentially infected tissues overlying the spinal defect (Mayfield 1981). The contents of the spinal canal are not excised. In paralytic scoliosis there is often more bleeding during the surgery because the muscles are weak and fibrotic and do not contract so aiding vasoconstriction. The venous canals in the osteoporotic bone are also larger and bleed more. In addition, the circulating blood volume is smaller in patients with a reduced muscle bulk and as a result they are less tolerant of excessive blood loss.

A very long fusion is usually necessary and there may be insufficient bone graft material from the iliac crests which are frequently hypo-

plastic and osteoporotic. Bone may be taken from both iliac crests and additional homologous bone may often be necessary. The author has found that deep frozen degenerate femoral heads removed during total hip replacements are a satisfactory source of homologous bone.

The spine is exposed by a meticulous subperiosteal dissection with removal of all remnants of soft tissue from the posterior bony elements. An interfacetal intertransverse fusion (Moe 1972) is performed followed by a deep and thorough decortication of all the posterior bony structures from the midline out to the tips of the transverse processes. This is followed by the application, throughout the fusion area, of large amounts of bone graft material cut into matchsticks. In myelomeningocele patients, the large

lumbar laminar defect and hypoplastic interfacetal joints make posterior fusion difficult and the incidence of pseudarthrosis may be as high as 46% (Osebold *et al.* 1982). In order to minimize this problem large amounts of graft material are laid bilaterally in the paraspinal gutters, on the sides of the defective laminae and on top of the vestigial transverse processes from the lower thoracic region to the sacrum.

Angular kyphotic curves in myelomeningocele patients are very rigid and it is necessary to first osteotomize the spine and resect a number of the vertebral bodies in order to gain correction. This is followed by instrumentation and posterior fusion (Fig. 7.3) from the thoracic region to the sacrum to prevent recurrence (Poitras & Hall 1974, Lindseth & Stelzer 1979).

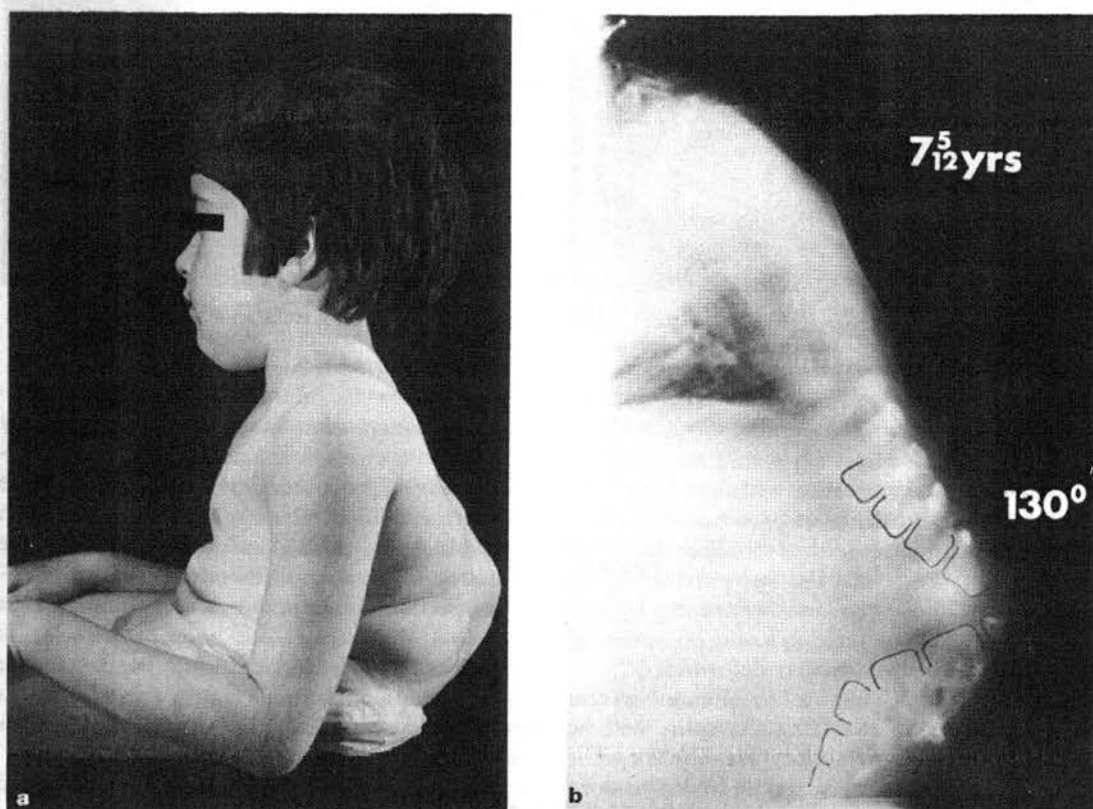


Fig. 7.3. (a) and (b) A seven-year-old girl with a myelomeningocele and a 130° angular kyphus in the lumbar region.

Indications

Most neuromuscular scoliotic curves remain fairly mobile until a late stage and a posterior spinal fusion with Harrington instrumentation is often sufficient providing the posterior elements are intact and there is not a very severe rigid curve extending to the sacrum. In myelomeningocele patients where there is a large laminar defect and insufficient posterior bony structures to fuse satisfactorily, it is usually necessary to supplement the posterior fusion by an anterior fusion with Dwyer instrumentation.

Contraindications

There are no contraindications to the use of Harrington instrumentation but the technique has certain disadvantages. The fixation of the hooks to the bone is not sufficiently secure to do without external immobilization. An underarm plaster jacket or brace is therefore necessary until the spine is solidly fused and this usually requires nine months. A plaster jacket can cause pressure sores if there is a sensory deficit and a removable type of plastic jacket is preferable.

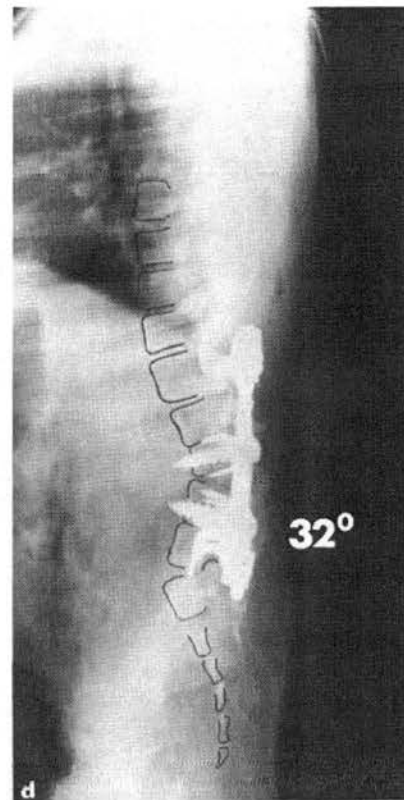


Fig. 7.3. (c) and (d) The deformity has been corrected to 32° by resecting the vertebrae at the apex of the kyphus. A posterior spinal fusion has been performed and correction maintained by a combination of Harrington and Dwyer instrumentation.

Anterior fusion with Dwyer instrumentation

In the late 1960s, Dwyer of Sydney, Australia, devised a completely different technique for spinal correction and internal fixation which was applied anteriorly to the vertebral bodies. Dwyer initially thought his technique was best applied to idiopathic scoliosis (Dwyer 1973), but the method is now mainly used in combination with a posterior fusion and Harrington instrumentation to correct severe neuropathic curves.

Technique

The convex side of the scoliosis is exposed through an extensive trans-thoracic and retroperitoneal approach in which the dia-

phragm is divided circumferentially. The intervertebral discs and vertebral end plates are excised throughout the length of the curve and the spaces packed with bone chips from an excised rib. Starting at one end of the curve, specially designed screw-and-staple units are inserted into each vertebral body and a flexible, braided titanium wire cable is passed through the holes in the heads of the screws. Adjacent vertebrae are compressed and the tension is maintained by crimping the screw heads into the cable. The process is repeated throughout the length of the curve which is gradually straightened (Fig. 7.4[a], [b] and [c]). Zielke *et al.* (1978) have modified the technique in an attempt to correct vertebral rotation as well as the lateral curvature. They replaced the Dwyer cable with a solid rod passed through the heads of the screws which are placed to derotate the

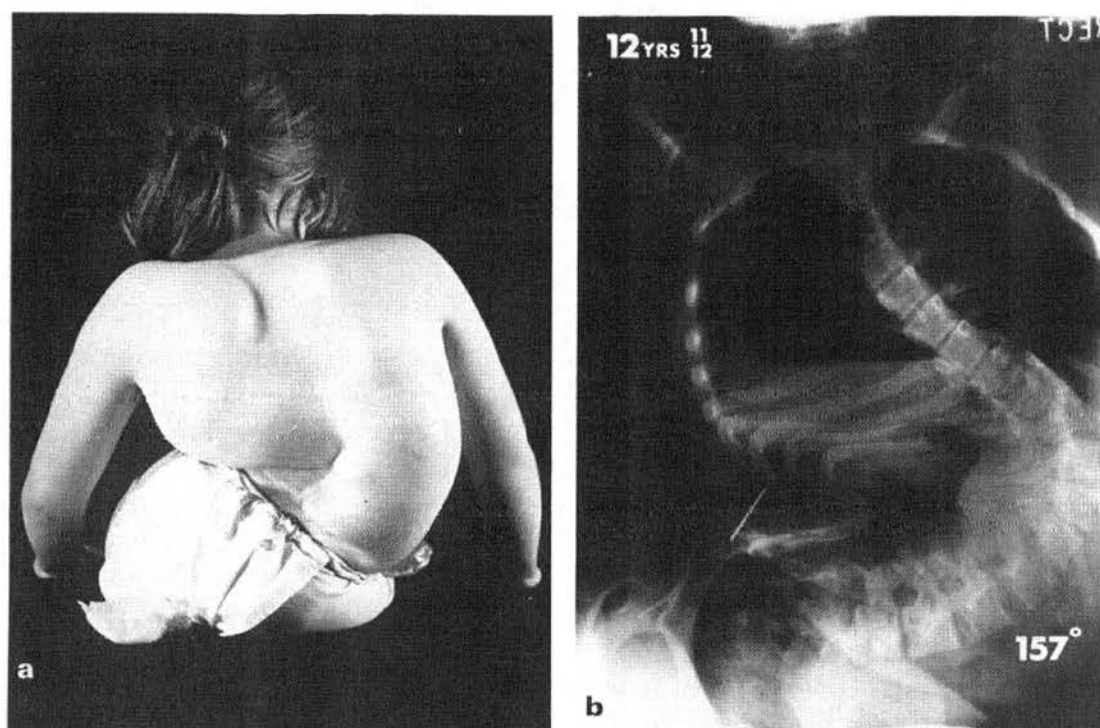


Fig. 7.4. (a) and (b) A twelve-year-old girl with a myelomeningocele and a very severe collapsing thoracolumbar lordoscoliosis measuring 157°. There is severe pelvic obliquity; she sits with all of her weight on one buttock and requires both arms for support.



Fig. 7.4. (c) The curve has been corrected to 67° by Dwyer instrumentation and fused anteriorly.

vertebral bodies as compression is applied (Moe *et al.* 1983).

Using these methods of anterior instrumentation an excellent degree of correction can be obtained and the segmental fixation is much more secure than with a Harrington distraction rod which grasps only the two end vertebrae. However, like Harrington instrumentation the internal fixation is not sufficiently secure to do without external immobilization until the fusion is solid. The major disadvantage of Dwyer instrumentation is that, for anatomical reasons, the screws cannot be easily inserted above the sixth thoracic vertebra or into the sacrum. Unfortunately neuromuscular collapsing curves are usually very long and, therefore, it is not always possible to correct and fuse the full extent of the deformity through an anterior approach. The typical thoracolumbar lordoscoliosis can only be satisfactorily fused and instrumented anteriorly from the tenth or eleventh thoracic

vertebra to the fourth or fifth lumbar vertebra. A second disadvantage is that although the Dwyer technique provides good correction of the scoliosis, the incidence of pseudarthrosis is much higher than with Harrington instrumentation and posterior fusion alone. Bonnett *et al.* (1976) found that in patients with cerebral palsy the incidence of pseudarthrosis with Dwyer instrumentation alone was 72% compared with 40% with Harrington instrumentation alone. For these reasons, anterior instrumentation and fusion is never sufficient and always requires to be supplemented by a second stage posterior fusion with Harrington or Luque instrumentation usually carried out 10–14 days after the anterior surgery (Fig. 7.4[d] and [e]). This provides even greater correction because the spine



Fig. 7.4. (d) Using Harrington instrumentation, further correction to 43° has been obtained and the spine has been fused posteriorly from the upper thoracic region to the sacrum.

has been released both anteriorly and posteriorly. It also allows extension of the fusion, if necessary, to the upper thoracic region and distally to the sacrum to control the pelvic obliquity which is present in collapsing curves and must be included in the fusion (O'Brien *et al.* 1975).



Fig. 7.4. (e) One year after the surgery, the patient has a balanced spine over a level pelvis and can sit without support.

The combination of an anterior spinal fusion with Dwyer instrumentation and a posterior fusion with Harrington instrumentation produces an excellent degree of correction with a very low incidence of pseudarthrosis and a stable spine (McMaster 1986). These results are much better than when Harrington instrumentation is used alone. Leong *et al.* (1981) found that in patients with poliomyelitis the combined procedure usually corrected collapsing thoracolumbar curves by over 60% whereas Harrington instrumentation alone usually achieved less than 40% correction. The incidence of pseudarthrosis was also reduced from 25% with

Harrington instrumentation alone to 12.5% with the combined procedure. Lonstein and Akbarnia (1983) found similar results in patients with cerebral palsy. The degree of correction was improved from 55% with Harrington instrumentation alone to 70% by the combined procedure and the incidence of pseudarthrosis lowered from 22% to 5.9%. Osebold *et al.* (1982) found that in patients with myelomeningocele the degree of correction was improved from 46% to 62% by the combined procedure and the incidence of pseudarthrosis lowered from 46% to 23%.

Indications

The main indication for an anterior spinal fusion with Dwyer instrumentation followed by a posterior spinal fusion and instrumentation is a severe rigid thoracolumbar or lumbar scoliosis with severe pelvic obliquity especially when combined with a lordosis. In these patients it is essential to level the pelvis and this can only be achieved by the combined procedure (O'Brien *et al.* 1975). The second indication is in patients with a myelomeningocele where there is a deficiency of the posterior elements and posterior fusion with Harrington instrumentation alone is likely to fail (Sriram *et al.* 1972).

Contraindications

The presence of a kyphosis is a contraindication to Dwyer instrumentation because the screws in the vertebral bodies lie anterior to the axis of flexion of the spine and when the cable is tightened it causes the kyphosis to increase. In some patients there is a severe rotation of the vertebrae in the scoliotic curve and this gives the appearance of an associated kyphosis (Stagnara *et al.* 1978). Correct placement of the screws during instrumentation will cause derotation of these vertebrae as the scoliosis is corrected with reduction in the rotatory kyphosis (Lonstein & Winter 1982). This type of kyphosing scoliosis is particularly suitable for the Zielke instrumentation.

Anterior fusion is usually contraindicated in patients with Duchenne muscular dystrophy and spinal muscular atrophy because they usually have severely impaired respiratory function, and the transthoracic approach necessitates dividing the diaphragm which is the main respiratory muscle and adds considerably to the operative risks.

It may not be possible to insert the Dwyer screws into the vertebrae of very young children because once the end plates have been removed, there is insufficient bone to hold the screws. In these young children it is possible to perform an anterior release and fusion without Dwyer instrumentation and follow this in the usual way with a second stage posterior spinal fusion with instrumentation.

Segmental spinal instrumentation (Luque technique)

Posterior spinal fusion with Harrington instrumentation when used alone or in combination with anterior fusion and Dwyer

instrumentation has been used successfully in the treatment of neuromuscular spinal deformities but has proved to be less than ideal. The main disadvantage of these methods used either singly or in combination is that they do not provide sufficiently secure internal fixation and additional support is necessary in a plaster jacket or brace for at least nine months to allow the fusion to mature and the spine to become stable. This external support may further restrict an already impaired respiratory system and produce pressure sores in patients with anaesthetic skin. The ideal intraoperative method of correction and internal fixation for neuromuscular curves should be easily applied to a wide variety and combination of spinal deformities and provide sufficiently secure internal fixation to maintain correction without external support.

In 1973, Dr Eduardo Luque of Mexico City, began experimenting with wires passed beneath the laminae and around a Harrington distraction rod in an attempt to gain more rigid internal fixation of the spine in young patients with poliomyelitis. In 1975 after many modifications he finally introduced a totally different system of

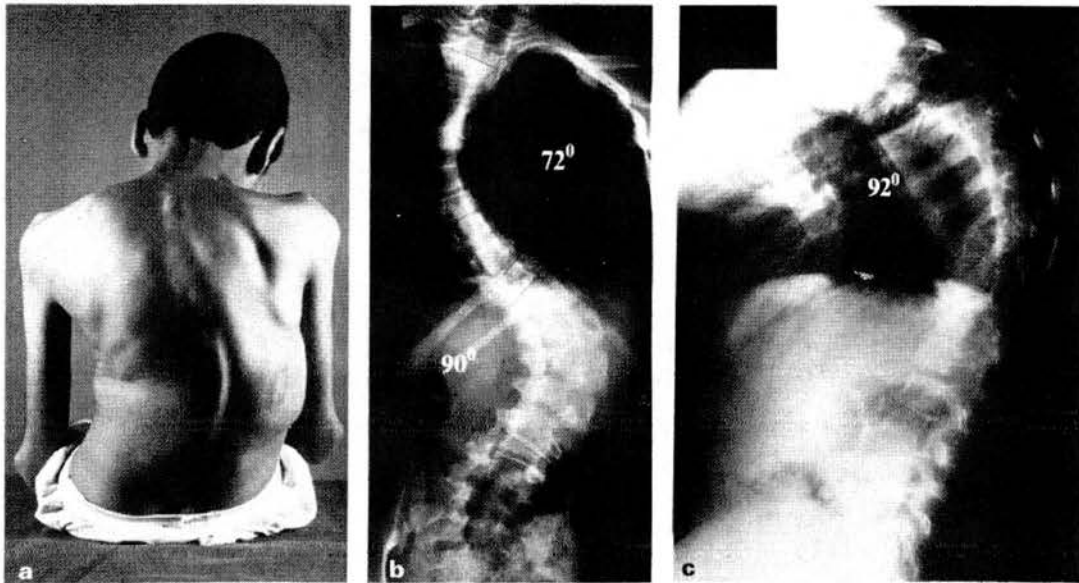


Fig. 7.5. (a), (b) and (c) A twelve-year-old boy with poliomyelitis and a severe collapsing thoracolumbar kyphoscoliosis.

spinal instrumentation in which the correcting forces are applied transversely to the spine at every level throughout the deformity rather than a distraction force across the concavity as in Harrington instrumentation or a compression force around the convexity as in Dwyer or Zielke instrumentation. The Luque method of segmental spine instrumentation also allows the deformity to be corrected in two planes as well as maintaining the normal physiological sagittal curves (Luque & Cardoso 1977, Luque 1982b). Because the technique provides an extremely rigid fixation, it eliminates the need for external support and is particularly applicable to all types of neuromuscular spinal deformities (Fig. 7.5). Initially the technique was used without fusion in an attempt to allow continued growth of the spine but eventually there was a high incidence of instrument failure and the method is now combined with a posterior fusion (Luque 1982a).

Technique

The spinal deformity is corrected and internally fixed by applying two prebent stainless steel rods, one on either side of the laminae, at every level throughout the length of the deformity. The two rods are bent to conform to the desired shape of the spine in both the sagittal and coronal planes as shown on the anteroposterior and lateral radiographs of the spine taken either with traction applied or the spine bent into a corrected position. These rods are attached to the spine at every level by means of wires passed beneath each side of the lamina and around each rod. The rods are bent at one end to an L shape to prevent migration up and down the spine. If pelvic fixation is required the short limb of the L is modified and driven into the transverse bar of the ilium on each side (Allen & Ferguson 1984). The spine is corrected as the contoured rigid rods are pushed into place and the wires are tightened

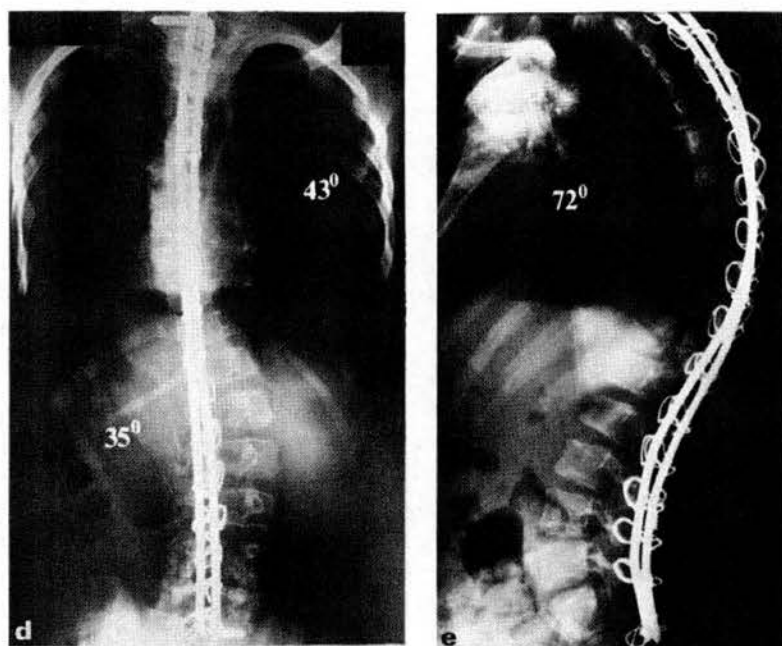


Fig. 7.5. (d) and (e) Using the Luque technique of posterior spinal segmental instrumentation, the spine has been well corrected and balanced over a level pelvis. The posterior spinal fusion extends from the cervicothoracic junction to the sacrum.

at each vertebral level by twisting. In this technique there is no distraction of the spine and correction is obtained in both the sagittal and coronal planes by a system of leverage and pushing and pulling the vertebrae into position. The system is further strengthened by wiring the rods together at several levels. A difficulty in this technique is predicting the amount of correction that can be obtained by prebending the rods. The primary objective is to balance the spine and it is important not to attempt over correction as this could be dangerous. At the end of the procedure the contoured rods should lie firmly against the laminae on either side of the base of the spinous processes. The two sites of attachment at each vertebral level provide an extremely rigid fixation and eliminates the need for external support (Fig. 7.5[f]). There is also the safeguard that if fixation is lost at any level it will not compromise the overall stability of the spine. In myelomeningocele patients there is difficulty in obtaining satisfactory fixation because of the

laminar defect and the wires are passed around the pedicles and fastened over the rods which lie in the lateral paraspinal gutters.

As with Harrington instrumentation, the success of Luque instrumentation ultimately depends on producing a solid stable posterior fusion extending over all of the instrumented levels. There is no evidence that rigid internal fixation alone will result in fusion of the spine. The technique of posterior fusion used with Luque rods is slightly different to that described earlier with Harrington instrumentation. It is not possible to decorticate the laminae or to carry out a very extensive excision of the facet joints because this could weaken the laminae or allow the wires to cut through. Large amounts of autogenous bone grafts are therefore placed along the gutters lateral to the rods and this provides the best chance for a massive fusion.

Indications

The Luque technique can be applied to all types and combinations of spinal deformities providing they are still relatively flexible as shown on the bending or traction spinal radiographs. However, the technique does not always provide as good a correction of large curves or pelvic obliquity as with Harrington instrumentation (Taddonio 1982). The likely explanation of this is the lack of a distraction force and downward pressure on the high side of the pelvis. However, it has the advantage over Harrington instrumentation in that correction can be more easily obtained in two planes. Postoperatively the patient can be mobilized in a few days and because external support is unnecessary there is no restriction of respiratory function. This is particularly advantageous in patients with spinal muscular atrophy or Duchenne dystrophy who must be mobilized very rapidly after surgery to prevent an increase in their functional disability.

Contraindications

There are no specific contraindications to the use of Luque instrumentation in neuromuscular spi-

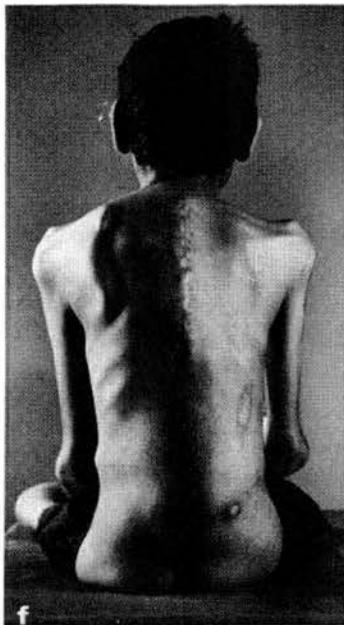


Fig. 7.5. (f) One week after the surgery, the patient is able to sit well balanced and without external immobilization.

nal deformities. However, in large rigid curves with severe pelvic obliquity, it may be necessary to first carry out an anterior spinal release with or without Dwyer instrumentation because Luque instrumentation alone would not gain sufficient correction (Fig. 7.6).

painful dysaesthesia which lasts for a few days. This is thought to be due to the movement of the wires against the nerve roots which occurs while the spine is being fused and before the wires are tightened.

The main disadvantage of the Luque techni-

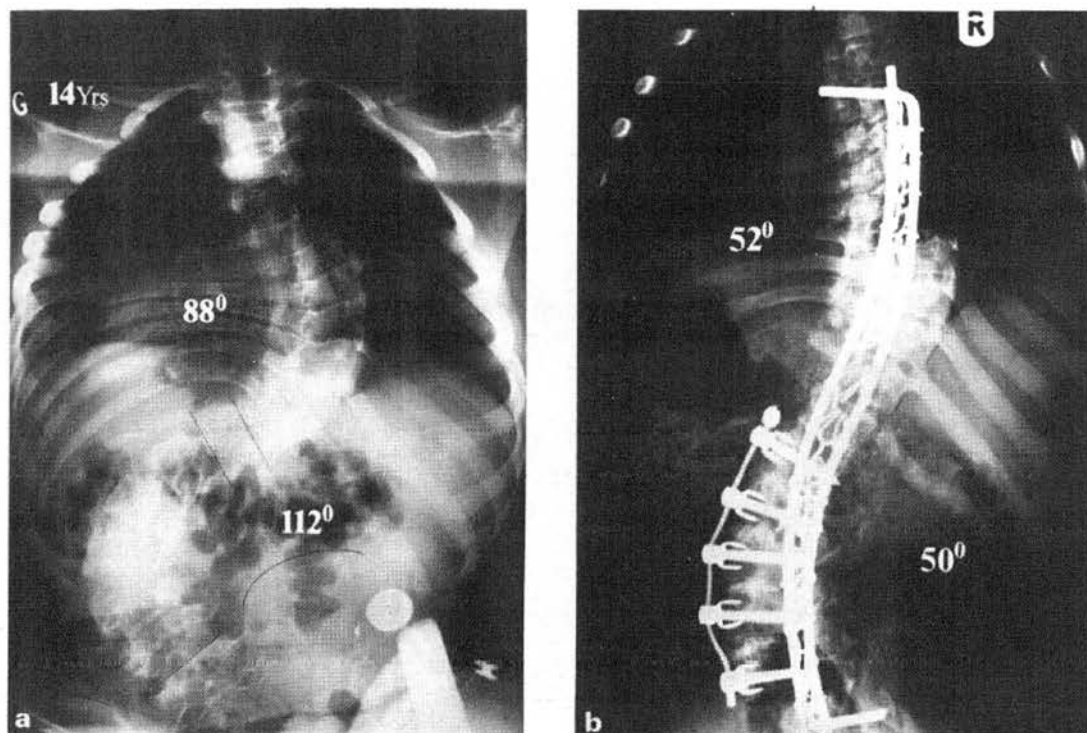


Fig. 7.6. (a) A fourteen-year-old boy with poliomyelitis and a very severe and rigid lumbar scoliosis with pelvic obliquity. (b) Because of the rigidity of the deformity, the lumbar scoliosis was corrected anteriorly by Dwyer instrumentation. Further correction was obtained ten days later using the Luque technique and the spine fused posteriorly from the upper thoracic region to the sacrum.

The Luque technique is a much more difficult and prolonged procedure than Harrington instrumentation and the desire to free the patient from external support must be weighed against the possible dangers of the technique. The main danger is that by passing the wire beneath the laminae the spinal cord could be damaged or an epidural haematoma could occur and press on the spinal cord. The most frequent complication is an isolated nerve root dysfunction producing

que is the large mass of metal that is applied to the posterior bony structures and decreases the area over which bone graft can be applied. This may interfere with the success of the posterior fusion and is particularly disadvantageous in the myelomeningocele patient who already has very little bone posteriorly. Radiographic assessment of the fusion is also difficult because of the overlying metal.

Postoperative management

Immediate care

Immediately after surgery the patient is nursed in a normal bed and log rolled from side to back to side. External support for the spine is unnecessary at this time provided internal fixation has been used. A Stryker turning frame is not used because many of these patients rely on diaphragmatic breathing and this is impaired when they are turned prone. Respiration may be helped by slightly elevating the end of the bed to prevent the abdominal contents from pressing on the diaphragm.

In the early stages, the most important aspects are to maintain respiratory function and to replace the blood loss. Care should be taken not to over transfuse these patients with fluids other than blood because it is possible to precipitate them into cardiorespiratory failure. If the preoperative respiratory function is poor it is possible that the patient will require supported ventilation in the postoperative period. Tracheostomy is rarely, if ever, required and an endotracheal tube with assisted positive pressure ventilation is nearly always sufficient. This is continued for a few days until the postoperative pain is gone and the patient has recovered sufficiently to breathe spontaneously.

Later care

After the immediate postoperative period, the aim should be to return these patients to their pre-operative status either walking or in a wheelchair by ten days. External support is unnecessary if Luque instrumentation has been used. However, if Harrington or Dwyer instrumentation has been used it will be necessary to apply some form of external support. The type of support will depend on the site of the curvature, skin sensation, pulmonary function and the reliability of the patient.

The most suitable form of postoperative external support is a bivalved total contact poly-

propylene body jacket similar to that worn in the conservative treatment of thoracolumbar and lumbar curves. This jacket is made from a mould of the patient taken after the surgical correction of the spinal deformity. The jacket which is light and comfortable allows regular inspection of the skin and this is particularly important in the myelomeningocele or spinal cord injured patient with impaired sensation. The jacket is only worn while the patient is upright and removed while in bed. A Milwaukee brace is used for upper thoracic curves as these would otherwise require a Risser-Cottrel type of plaster jacket. This type of brace is lighter than a cast and allows satisfactory chest expansion. The main disadvantage of both these types of brace is that they can be easily removed by the unreliable patient.

A plaster jacket of the lighter materials may only be used in the more mobile patients with normal sensation. An underarm jacket is sufficient for thoracolumbar and lumbar curves but a Risser-Cottrel jacket is necessary for higher curves. A plaster jacket should never be applied if there is anaesthetic skin or if it is likely to impair respiratory function. Some patients with weakness of their legs may find that the weight of the plaster makes walking more difficult and this is also a contraindication.

It is not necessary to include the hips in any form of postoperative spinal support as this will interfere with the mobility of the patient. The external support is worn for nine months to a year to allow the spinal fusion to mature and become stable.

Postoperative complications

The incidence of complications following the treatment of neuromuscular spinal deformities are much higher than for any other type of scoliosis. Death may occur in the postoperative period due to either cardiac or respiratory failure but the likelihood of this complication can be significantly reduced by careful pre-operative assessment and postoperative management. The problems of instrument failure and pseudarthrosis following surgery have already been dis-

cussed. Infection is always a problem and may affect either the chest or the wound. Failure to cough satisfactorily can result in postoperative atelectasis and bronchopneumonia. Wound infection is most frequent in the myelomeningocele patient with scarred skin and this can be reduced by plastic surgical procedures prior to the spinal fusion, avoiding entering the dura during surgery, and giving prophylactic antibiotics. Using prophylactic antibiotics, Osebold *et al.* (1982) reduced wound infection in myelomeningocele patients from 35 to 17%. Pressure sores can also be reduced by the use of removable jackets and regular inspection of the skin. Pathological fractures of the lower limbs due to osteoporosis can occur after prolonged postoperative bed rest and can be minimized by early mobilization.

Conclusion

We have now entered a new era in the surgical management of neuromuscular spinal deformities. The Luque method of posterior spinal segmental instrumentation promises to overcome many of the problems related to external immobilization and prolonged bed rest but longterm results are not yet available. Although this type of surgery is an arduous and complex task, it can be very rewarding to these severely disabled patients and even a relatively small gain in function can make a major difference to their lives. Many of these patients will spend most of their lives sitting and it is important that they should be able to do so with a balanced stable spine and have independent use of their hands for activities other than supporting themselves. The potential for disaster following surgery for neuromuscular spinal deformities is high, but satisfactory results can be achieved by expert surgical treatment in a specialized centre with adequate back-up facilities.

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Section G

Miscellaneous

SPINAL DEFORMITY IN EHLERS-DANLOS SYNDROME

FIVE PATIENTS TREATED BY SPINAL FUSION

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Five patients with classical Ehlers-Danlos syndrome developed severe spinal deformities. Two were shown to have type-VI collagen abnormalities. Three had a double structural scoliosis of the thoracic and lumbar regions, one had a single thoracic scoliosis and one had a thoracic kyphosis.

The curves first developed before the age of four years, and were not controlled by bracing. Major corrective surgery with posterior fusion was performed at a mean age of 11 years 8 months. Excessive blood loss could be controlled and although wound haematoma and dehiscence were common, they did not provide major problems. The spinal fusions healed satisfactorily.

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Ehlers-Danlos syndrome (EDS) is a rare hereditary disorder of collagen metabolism which has recently been classified into subgroups on the basis of the underlying defect in collagen fibril formation. The characteristic clinical features of all types of EDS are hypermobility of joints, which may sublux or dislocate, hyperextensibility of the skin and general friability of the soft tissues which are easily bruised and heal poorly with 'tissue-paper' scars.

Spinal deformity may occur in EDS, but very little is known about its type and incidence (Beighton and Horan 1969). Leatherman and Dickson (1988) stated that patients with EDS are more likely to develop spinal deformities, but that there was little evidence to suggest that these could become severe; surgical treatment should not be advised.

This paper describes the patterns of spinal curvature in five patients with EDS and reports the results of surgical treatment.

PATIENTS

From 1976 to 1991, five patients with classical EDS presented with spinal deformities to the Scoliosis Clinic at the Princess Margaret Rose Orthopaedic Hospital, Edinburgh. All were girls with hypermobile joints, severe ligamentous laxity, and hyperextensible skin. They all had a tendency to prolonged bleeding and evidence of poor soft-tissue healing with tissue-paper scarring.

The syndrome had been diagnosed between the ages of one and nine years as a result of recurrent subluxation or dislocation of shoulders, hips, sternoclavicular or patellofemoral joints. Since tissue typing became available the last two patients (cases 4 and 5) have been diagnosed as having type-VI collagen disorders. They were sisters, but there was no relevant family history in the other three patients.

Spinal deformity had first been recognised before the age of 4 years. Three patients were treated in braces, but the curves deteriorated and all five patients required major corrective spinal surgery at a mean age of 11 years 9 months (10 years 5 months to 13 years 7 months). All had posterior spinal fusion performed by the author, two with Harrington instrumentation and three with Luque L rods.

Case 1. A girl presented shortly after birth with Erb's palsy, hyperextensible skin, general ligamentous laxity and dislocatable hips and shoulders. A spinal deformity was noted, but was not treated until the age of 2 years 6 months, when she had an 89° smooth thoracic kyphosis extending from T3 to L3 with its apex at T9. A Milwaukee brace was worn until the age of 10 years 4 months when the kyphosis measured 45°. The patient then refused to wear the brace and the kyphosis deteriorated rapidly to 95° at the age of 13 years 7 months. She also developed poor respiratory function with a vital capacity of less than 50%.

A posterior spinal fusion was performed with Luque L-rod instrumentation extending from T1 to L2 and correcting the kyphosis to 41°. Postoperatively, an underarm plaster jacket was used for three months, then removed because of a pressure sore in the fragile skin over her kyphosis. At the age of 14 years 8 months she became increasingly dyspnoeic, her general condition deteriorated and she died of cardiorespiratory failure. Post-mortem examination revealed an empyema of the

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right lung and infarction of the left lung. The spinal fusion had healed satisfactorily.

Case 2. A girl known to have a scoliosis at the age of three years, but untreated, was diagnosed in Edinburgh at the age of nine years as a case of EDS with a major left thoracolumbar scoliosis from T11 to L4 measuring 97°, and a minor right thoracic scoliosis from T3 to T11 measuring 66°. The vertebrae at the apex of the thoracolumbar curve were markedly rotated resulting in an apparent kyphosis from T7 to L4 of 55°.

A Milwaukee brace failed to control the deformity and by the age of 11 years the thoracolumbar scoliosis had deteriorated to 115°, and the thoracic scoliosis to 83°, with an apparent kyphosis of 77°.

Posterior spinal fusion from T4 to L5, using a single Harrington distraction rod, corrected the thoracolumbar scoliosis to 75°, the thoracic scoliosis to 57° and the apparent kyphosis to 24°. Fusion used autogenous iliac-crest bone supplemented by allografts from a degenerate femoral head. An underarm plaster jacket was worn for six months, and the solid fusion showed no loss in correction at three years' follow-up.

Case 3. A girl, first seen at the age of three years after a shoulder dislocation, was diagnosed as having classical EDS. She also had a 60° left thoracic scoliosis from T6 to L1 with its apex at T9 (Fig. 1). A Milwaukee brace was worn intermittently and by the age of 11 years 9 months, the scoliosis had deteriorated to 100° (Figs 2 and 3) with marked rotation at the apex giving the appearance of a thoracic kyphosis of 117°. Respiratory function was moderately impaired.

A posterior spinal fusion from T6 to L2, using a single Harrington distraction rod, corrected the scoliosis to 73°. The postoperative course was complicated by respiratory distress requiring assisted ventilation for 12 hours. An underarm plaster jacket was worn until solid fusion had occurred at six months (Figs 4 and 5). By the age of 16 years the scoliosis measured 74°.

Case 4. A girl diagnosed as having EDS at the age of three years also had a mild thoracolumbar kyphosis. No treatment was given and by the age of 11 years 9 months she had a left thoracolumbar scoliosis from T8 to L1 of 75° (Figs 6 and 7). Rotation resulted in an apparent kyphosis of 49° (Fig. 8).

Posterior spinal fusion was performed with Luque L-rod instrumentation extending from T4 to L4. Bone grafts from the iliac crest were supplemented by allografts from a degenerate femoral head. The thoracolumbar scoliosis was reduced to 15° and the sagittal profile returned to normal (Figs 9 to 11). An underarm jacket was worn for three months. A solid fusion developed and after 18 months the scoliosis measured 16°.

Case 5. The sister of case 4 was diagnosed as having EDS in early infancy. By the age of 1 year 5 months she had a 72° thoracolumbar kyphosis from T6 to L3. The kyphosis improved despite lack of treatment, but by the age of 10 years 5 months she had a double structural scoliosis: a

right thoracic curve of 68° extended from T4 to T11 and a left lumbar curve from T11 to L4 measured 50°. There was an apparent kyphosis at the junction of the curves.

Posterior spinal fusion using Luque L-rod instrumentation from T3 to L4 corrected the deformity; autografts and allografts were used as before. The thoracic scoliosis was corrected to 22°, the lumbar scoliosis to 18° and the sagittal profile to normal. An underarm jacket for three months allowed solid fusion with thoracic and lumbar curves of 38° and 16° respectively at one year.

Blood loss and wound healing. There was excessive capillary oozing at operation in all five patients, but this was controlled by hypotensive anaesthesia (Malcolm-Smith and McMaster 1983), local injection of 1:400 000 adrenaline solution, careful dissection using electrocautery, and wound packing. The mean perioperative blood loss was 1243 ml (1210 to 2140).

All five patients had wound haematoma, managed successfully by repeated aspiration and pressure dressings. There was wound dehiscence in two patients about two weeks after surgery; in both cases it healed satisfactorily without infection after secondary suture. All the wounds healed with tissue-paper scarring.

DISCUSSION

Some patients with EDS develop progressive spinal deformities at a relatively early age. All the curves reported here developed before the age of four years, and were severe by 11 years. Two patients (cases 1 and 3) had severe thoracic deformities causing respiratory impairment.

The prevalence of spinal deformity in EDS is uncertain. Beighton and Horan (1969) reported that 18 of 100 EDS patients had a scoliosis, but did not describe the pattern and severity of deformities. Of the 27 children under the age of 15 years in their series, only two had mild scoliosis, and another three children had a mild kyphosis due to slight anterior thoracolumbar wedging. Similar wedging has been described by Macfarlane (1959) and Coventry (1961) but was not seen in the present series.

It has been suggested that spinal deformity in EDS is more commonly associated with type-VI collagen abnormality, due to deficiency of the enzyme lysyl hydroxylase. Sussman et al (1974) reported one patient with type-VI EDS who presented as a teenager with a 45° thoracic scoliosis, which had progressed with no treatment to 80° by the age of 49 years. Patients with type-VI EDS are also more likely to have ophthalmic complications such as rupture of the globe and retinal detachment (Pinnell et al 1972; Sussman et al 1974). Tissue typing was performed only in cases 4 and 5; both were type-VI, but neither patient had eye problems.

All five spinal deformities were difficult to control in a brace and deteriorated rapidly during the adolescent growth spurt after the age of ten years. It is likely that lax



Fig. 1

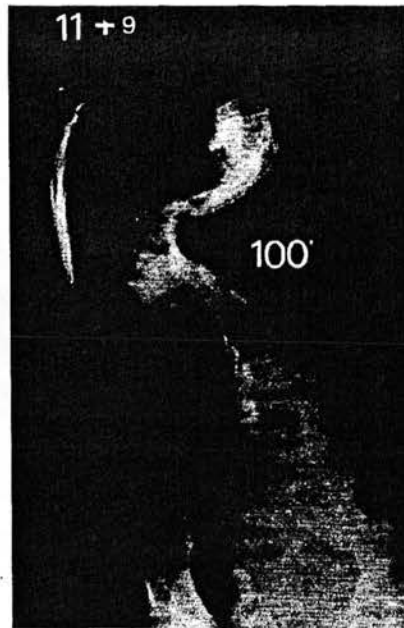


Fig. 2



Fig. 3

Case 3. Figure 1 – Radiograph of a girl aged 3 years 1 month showing a 60° left thoracic scoliosis. Figures 2 and 3 – By the age of 11 years 9 months, the scoliosis had deteriorated to 100°. The thoracic cage was deformed and respiratory function was impaired. Figures 4 and 5 – After posterior spinal fusion from T6 to L2, with Harrington instrumentation, the scoliosis was reduced to 73° at the age of 12 years 7 months.



Fig. 4



Fig. 5

ligaments were unable to maintain spinal balance in the presence of rapid longitudinal growth.

Four of the five developed a significant scoliosis and all these curves deteriorated rapidly at the onset of the adolescent growth spurt, requiring surgery at a mean age of 11 years for major curves of a mean size of 88° (66 to 115). It is probable that without such treatment, these curves would have rapidly become worse.

Little has been reported of the surgical management of spinal deformity in EDS. Leatherman and Dickson (1988) consider that surgery should be avoided, presumably because of the risks of bleeding and poor healing of

soft tissues, but give no supporting information. This series shows that, with care, extensive posterior spinal fusion and correction with instrumentation is possible, but that there may be complications. A number of different techniques were used; all provided an acceptable improvement in the frontal plane (mean 58%; range 35% to 78%). All the scoliotic curves, however, had a significant rotation producing an apparent kyphosis; this required segmental instrumentation to correct sagittal alignment (see Figs 8 and 11).

During surgery, bleeding was controlled by careful technique and hypotensive anaesthesia, and wound hae-

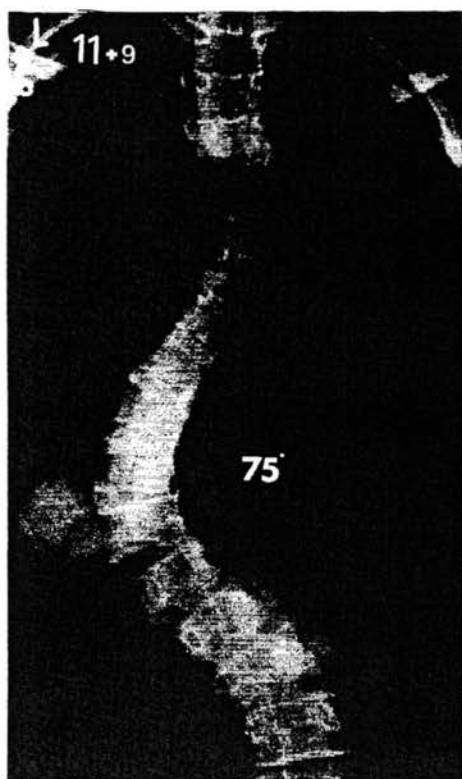


Fig. 6

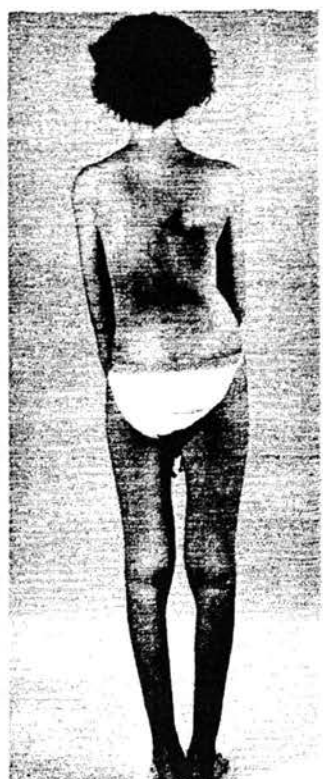


Fig. 7



Fig. 8

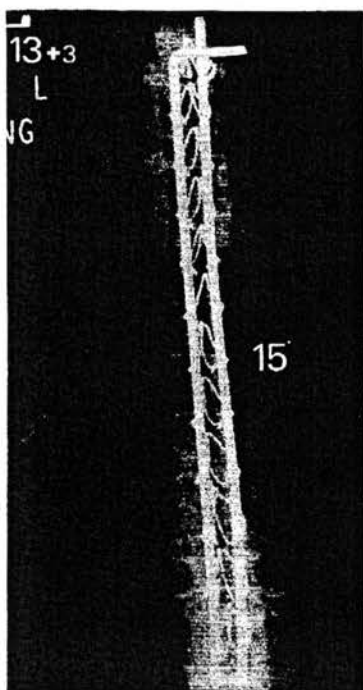


Fig. 9



Fig. 10



Fig. 11

Case 4. Figures 6 and 7 – Radiograph and photograph of a girl aged 11 years 9 months with a 75° left thoracolumbar scoliosis. Figure 8 – A lateral view shows marked rotation of the scoliosis producing a 49° pseudokypnosis. Figures 9 to 11 – After a posterior spinal fusion from T4 to L4, with Luque L-rod instrumentation, the scoliosis was reduced to 15° and sagittal alignment had been returned to normal.

matomas were controlled by repeated aspiration. Wound dehiscence was also common but was usually superficial and healed satisfactorily after secondary suture. The bone tended to be slightly osteoporotic, but there was no apparent delay or failure in bone healing. Both allografts and iliac-crest autografts were used; all patients developed a solid and stable fusion.

Severe spinal deformities may develop relatively early in patients with EDS. Major corrective spinal surgery may be needed and is possible with careful management of bleeding and soft-tissue problems. Spinal fusions heal satisfactorily and provide stable correction.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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Surgical Correction of Developmental Scoliosis Following Cardiac Transplantation

Case Report

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Michael J. McMaster, MD, FRCS,* and Mary Rose, FRCA†

Study Design. Case report.

Objective. To present a pediatric patient who underwent successful 2-stage anterior and posterior scoliosis surgery 20 months after cardiac transplantation.

Summary of Background Data. Cardiac transplantation has increased the life expectancy in children with end-stage cardiac failure caused by congenital heart disease or cardiomyopathy. Scoliosis is commonly associated with congenital cardiac disease. Previous reports have suggested that anterior scoliosis surgery is contraindicated after heart transplantation.

Methods. We describe the case of a 13-year-old patient who underwent staged anterior and posterior spinal arthrodesis to correct a severe thoracic developmental scoliosis.

Results. The 2 procedures were performed 1 week apart. Following the first stage, pulmonary edema had developed, which required reintubation and administration of diuretics. Significant blood loss occurred during the posterior spinal fusion. The patient had a satisfactory correction of her scoliosis and made a good recovery. Three years after surgery, she had no complaints of her back and had a solid spinal fusion.

Conclusions. Elective anterior and posterior spinal arthrodesis can be safely performed in cardiac transplant recipients with severe developmental scoliosis. This procedure can achieve optimum correction of the curvature and a successful outcome without long-term medical or technical complications.

Key words: cardiac transplantation, cardiopathy, developmental scoliosis, spinal surgery, immunosuppressive agents, risk assessment. *Spine* 2006;31:E713-E717

In 1967, Christian Barnard¹ was the first to perform successful cardiac transplantation on a patient with end-stage cardiac failure using the heart of a road accident victim. Following more recent improvements in immunosuppression, which have reduced the potential complication of allograft rejection, this operation has been

routinely performed in pediatric patients in many institutions since 1982.² The indications include primarily congenital cardiac disease and less commonly severe cardiomyopathy. As heart transplantation becomes an accepted therapeutic treatment, the population of pediatric recipients is increasing, and their life expectancy is significantly improved.³⁻⁵ A previous study by Lawrence and Fricker² has shown that children can adapt successfully to the major physical and emotional assault of such an extensive procedure. In addition, cardiac transplantation dramatically affects the quality of life of the surviving children by allowing them to function at a level that is appropriate for their age.

While the long-term survivorship of pediatric heart transplant patients increases, the possibility of these patients having other pathologies develop that require surgical treatment is also growing. There are published reports concerning this group of patients undergoing cholecystectomy, orthopedic, thoracic, or vascular operations, flap advancement for sternal wound infections, procedures of the colon and rectum, and repair of inguinal or incisional hernias with a significant risk of related complications and death.⁶⁻¹¹ The association of scoliosis with congenital cardiac disease is well established.¹²⁻¹⁷ Most cases involve developmental curvatures, resembling idiopathic scoliosis. There is usually no evidence of congenital abnormalities in the formation of the vertebral column. In 2001, Ceroni *et al*¹⁸ were the first to report a 15-year-old patient with end-stage cardiac failure who underwent posterior spinal arthrodesis with a good outcome 14 months after heart transplant. In this article, the investigators suggested that an anterior approach to the spine is contraindicated to avoid reentering the previously operated pleural cavity and that a lesser degree of curve correction using a posterior-only procedure should be accepted.¹⁸ The purpose of this report is to present our experience in successfully performing staged anterior and posterior spinal fusion on a pediatric patient with end-stage cardiac failure and developmental scoliosis 20 months following cardiac transplantation.

■ Case Report

A girl aged 10 years and 5 months presented to our spinal outpatient clinic with a scoliosis. Radiographs of the spine revealed a low right thoracic curve measuring 50°, which was producing a rib prominence adjacent to the convexity of the scoliosis as well as an elevation of the right shoulder. At the age of 2 years, our patient had been

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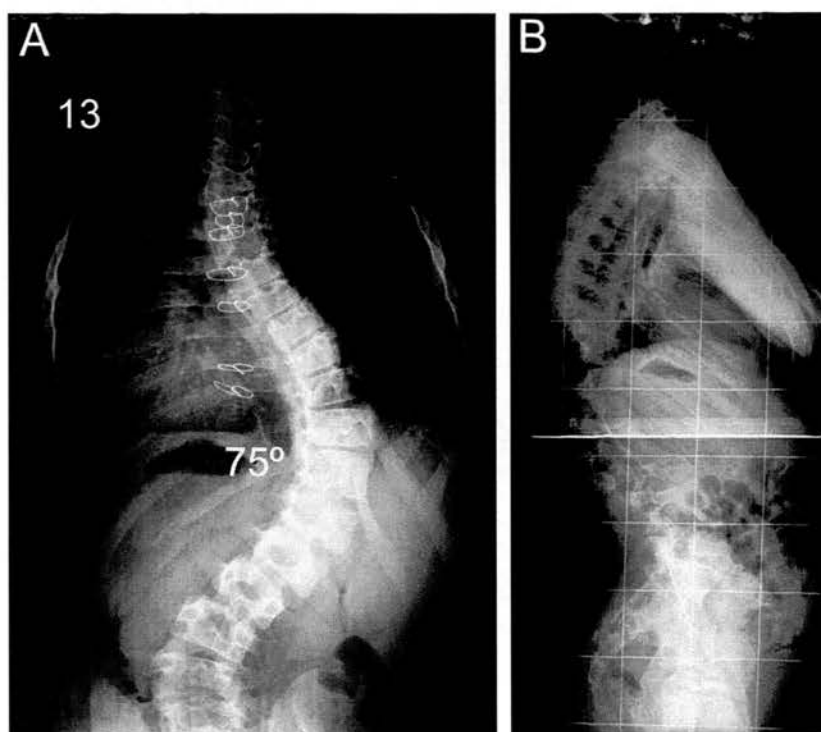


Figure 1. Preoperative anteroposterior (A) and lateral (B) radiographs of the spine at age 13 years showing a severe lower thoracic scoliosis measuring 75°, which caused listing of the trunk and translocation of the thoracic cage to the right, as well as an associated rotatory thoracolumbar kyphosis.

diagnosed as having an atypical hypertrophic spongiform cardiomyopathy with secondary pulmonary hypertension and severe exercise intolerance. This diagnosis gradually deteriorated to end-stage cardiac failure, and, at the time when she attended our clinic, she was awaiting a cardiac transplantation. We decided to treat her developing scoliosis with an underarm spinal brace until she would be medically fit to undergo surgical correction.

At the age of 11 years and 6 months, orthotopic domino heart transplantation was performed through a sternotomy with a satisfactory outcome, resulting in marked improvement of her cardiac function and overall physical fitness. After the operation, she was placed on oral immunosuppressive agents (azathioprine 100 mg once daily and tacrolimus 300 mg twice daily) but did not require corticosteroids. Following her cardiac transplant, acute renal failure developed, requiring hemofiltration, which recovered without any further problems. The spinal brace was discontinued during and after the cardiac operation, and the scoliosis subsequently progressed to 75° at the age of 13 years (Figure 1). Because of the fact that the patient was skeletally immature and the iliac apophyses had not yet appeared, increasing the risk of a crankshaft effect, a 2-stage anterior and posterior spinal fusion was performed 20 months following the heart transplant. A multidisciplinary review indicated that corrective anteroposterior surgery for her developmental scoliosis could be attempted.

Preoperative Assessment

A detailed cardiologic evaluation was performed, including echocardiography, which confirmed a structurally normal heart with good systolic and diastolic function, no residual pulmonary hypertension, and an ejection

fraction of 81%. A cardiac catheterization revealed normal coronary arteries. Respiratory function tests showed a uniform reduction in all lung volumes.

The cardiothoracic surgical team involved in her transplant was consulted and requested continuation of the immunosuppressive agents throughout the perioperative period (azathioprine 100 mg once daily and tacrolimus 300 mg twice daily). They also highlighted the need for Cytomegalovirus (CMV) negative blood if a transfusion was required, to avoid the risk of post-transplant CMV infection. The hematologists were involved and did not identify any adverse effects of the prolonged immunosuppressive therapy that could potentially affect her scoliosis surgery other than a chronic anemia (Hb: 9). To address the anemia, the patient was started on iron and folic acid tablets, and received 3 doses of erythropoietin 4500 IU before surgery, as well as 1 additional dose between the first and second stages of the procedure. Blood predonation was not performed.

A urologic assessment showed decreased creatinine clearance of 35% of predicted, which indicated that hypotensive anesthesia would not be safe during scoliosis surgery and that nonsteroidal antiinflammatory medication should not be used for postoperative pain control. The patient was markedly underweight (29.8 kg; 5 kg less than the third percentile for her age), and a detailed nutritional evaluation was performed to provide dietary advice, as well as nutritional supplements and optimize her for the surgery.

Spinal Procedure

The patient underwent a 2-stage anterior and posterior spinal arthrodesis. The senior author (M. J. M.) performed both stages. The first stage involved a right tho-

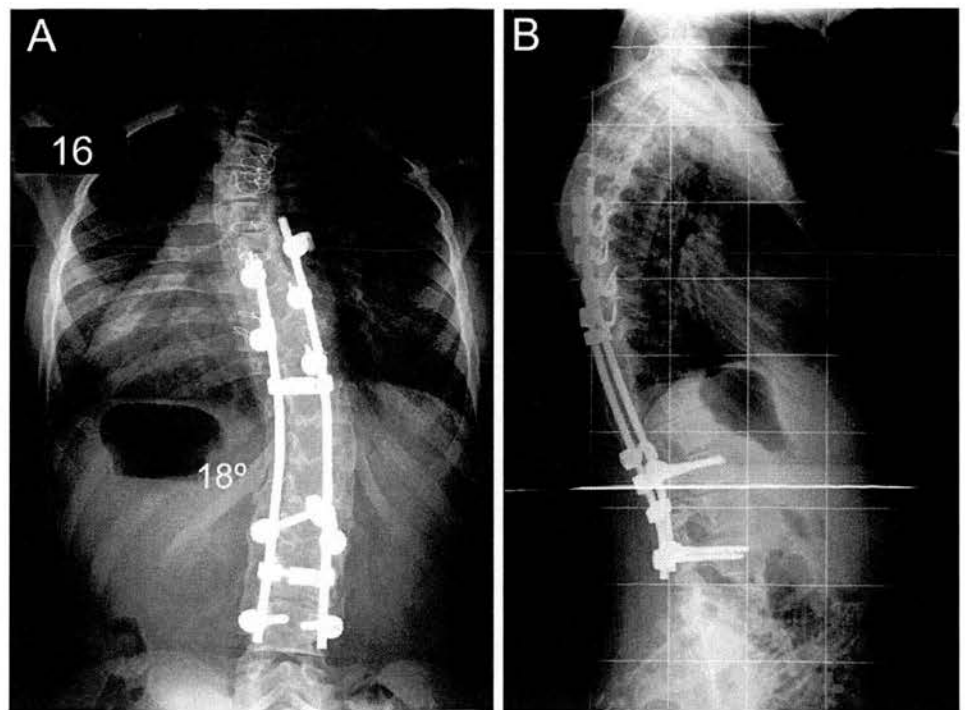


Figure 2. Anteroposterior (A) and lateral (B) radiographs of the spine at age 16 years, 3 years following 2-stage anterior and posterior spinal arthrodesis revealing correction of the lower thoracic scoliosis to 18°. Note the mild elevation of the left shoulder, which is caused by the development of an upper thoracic scoliosis.

racoabdominal transdiaphragmatic retroperitoneal approach to the spine to perform multiple discectomies from T9 to L2, with the aim to release the scoliosis and achieve anterior intervertebral fusion. Intraoperative blood loss was measured at 470 mL, and the patient was extubated at the end of the operation without problems. She was transferred to the intensive care unit and started on nasogastric feedings. On the first day following her surgery, respiratory distress developed because of fluid retention and pulmonary edema, which necessitated reintubation. This distress was treated with fluid restriction and administration of a diuretic (frusemide). She required transfusion with 2 U of de leukocytized CMV-negative blood.

One week later, the patient underwent a second-stage posterior spinal arthrodesis extending from T6 to L4, with the use of autologous bone graft (rib and iliac crest) and AO-Universal Spine System pediatric instrumentation (Stratec Medical, Oberdorf) secured to the spine by a combination of pedicle screws distally and hooks proximally. Intraoperative blood loss was measured at 1800 mL, and the patient required transfusion with 4 U of de leukocytized CMV-negative blood.

Continuous blood pressure monitoring by arterial line and central venous pressure monitoring were performed throughout both procedures, as well as cardiac recording through an electrocardiogram. Spinal cord monitoring was also obtained during both operations by recording somatosensory evoked potentials and did not indicate any neurologic deficit. Three doses of intravenous antibiotic prophylaxis, including cefuroxime for the first stage, and teicoplanin and ceftazidime for the second stage, at induction of anesthesia, at the end of the sur-

gery, and 6 hours after surgery were given for each procedure.

Postoperative Recovery

Following the second-stage posterior spinal fusion, the patient was electively ventilated for 3 days and made a good recovery after discharge from the intensive care unit on the third postoperative day. The immunosuppressive medications were continued throughout the perioperative period and after surgery, as indicated by the cardiothoracic team. She stood up for the first time on the fourth day without the need for an external support to the spine and was ready to leave hospital on the eighth postoperative day after a total period of hospitalization of 15 days.

Our patient made a good recovery, and there were no problems with wound healing. Her lower thoracic scoliosis was corrected very satisfactorily to 18° and has remained stable at follow-up. At the latest clinical visit, 3 years after surgery, she had no complaints of her back, and there was good radiographic evidence of a solid fusion across the instrumented levels (Figure 2). However, her left shoulder was slightly elevated because of the development of a small upper thoracic scoliosis, which in retrospect should have been included in the levels of the spinal arthrodesis during the index procedure.

Discussion

The incidence of scoliosis in patients with congenital cardiac disease has been reported to range between 4% in those patients who have not undergone operative treatment and 11% in children who have undergone heart surgery.^{15,17} This variation of incidence has led previous

investigators to assume that there may be an association between thoracotomies in patients with congenital cardiac disorders and the development of scoliosis.^{15,19} Most curves have their convexity toward the thoracotomy side.²⁰ However, other studies have failed to show a relation between the side of the thoracotomy and the convexity of the scoliosis.^{16,21,22} More recently, Ruiz-Iban *et al*²³ have shown that scoliosis can also develop following a median sternotomy.

There are 2 types of scoliosis that can be seen in conjunction with congenital heart disease: congenital and developmental scoliosis. Congenital scoliosis is caused by an asymmetrical failure of development of ≥ 1 vertebrae, resulting in a localized imbalance in the longitudinal growth of the spine and an increasing curvature, which continues to progress until skeletal maturity.^{24,25} Congenital cardiac disease may be found in 10% of patients with congenital scoliosis.¹⁶

Developmental scoliosis tends to present at an average age of 11 years, with convex left curves most commonly located in the proximal thoracic spine and convex right curvatures in the lower thoracic spine.^{13,15} There is no evidence of morphologic anomalies in the formation or segmentation of the vertebrae that could be the cause of the deformity. Despite recent advancements in surgical techniques and spinal instrumentation, corrective surgery to address scoliosis involves significant challenges and has the potential for serious complications. Cardiac transplant patients undergoing scoliosis surgery are at a considerably higher risk because of the altered physiology of the denervated heart as well as the adverse effects of immunosuppressive medication. This risk varies primarily according to the patient's preoperative cardiac function.²² Although orthopedic procedures are not contraindicated in this compromised patient population, special precautions should be taken to accommodate for the increased risks.⁶ To our knowledge, there has been only 1 previous report of a patient with cardiac transplantation undergoing subsequent scoliosis surgery, and this was with a posterior spinal arthrodesis alone.¹⁸

The transplanted heart does not respond acutely to hypovolemia or hypotension with a neural reflex tachycardia, as the native heart would normally do. In addition, anesthetic agents acting *via* the autonomic nervous system have no effect on the denervated cardiac muscle. The transplanted heart has an altered response to stress. It primarily responds by an increase in the cardiac output and stroke volume. This increase in cardiac output is mediated by the Frank-Starling mechanism and, therefore, depends on venous return. Therefore, resting heart rate is increased and will respond to circulating catecholamines (humoral reflex), but the response is delayed. Because the denervated heart is preload dependent, Ceroni *et al*¹⁸ have recommended continuous transesophageal echocardiography to provide information on cardiac filling during the spinal procedure. Other anesthetic concerns include the increased potential for respiratory complications caused by the previously per-

formed thoracotomy. Therefore, intraoperative management by experienced cardiopulmonary anesthetists and postoperative intensive care support are the prerequisites for successful corrective scoliosis surgery.

The immunosuppressed state of the cardiac transplant patient increases the susceptibility to infection following spinal surgery. If septic complications develop after such a major operation, these may threaten the patient's life.^{6,10,26,27} Delayed healing of the spinal wound may be anticipated, however, that did not occur in our patient or in the patient reported by Ceroni *et al*.¹⁸ Malignancies may also develop as the result of the chronic immunosuppression.⁶ Other orthopedic complications following heart transplantation include avascular necrosis of the hip, soft tissue infections, stress fractures, osteomyelitis, osteoporosis, and osteomalacia.^{28,29}

Immunosuppressive agents, especially corticosteroids and tacrolimus, can have a deleterious effect on bone mineral metabolism and create osteoporosis. In a previous study, Stempfle *et al*³⁰ have shown that a high-dose tacrolimus-based immunosuppressive regimen is associated with rapid bone loss early after cardiac transplantation. Beyond the first 6 months after heart transplant, calcium, vitamin D, and hormone supplementation in cases of hypogonadism lead sufficiently to bone mineral recovery.³⁰ Osteoporosis weakens the vertebral body and posterior bony arch, thus complicating instrumentation fixation and increasing blood loss during spinal arthrodesis. Therefore, the surgical technique may have to be modified to reduce blood loss and operating time without compromising the outcome of the procedure. The posterior stage of the spinal fusion in our patient was associated with increased blood loss, especially because hypotensive anesthesia was contraindicated. However, there were no problems with spinal fixation.

In patients who have undergone cardiac transplantation, repeat thoracotomy for scoliosis correction has previously been contraindicated.^{18,22} In addition, Bhatia *et al*⁶ have reported a mortality rate of 45% in 11 patients who underwent heart transplantation and required 17 repeat thoracic procedures. The increasing popularity of median sternotomy for cardiothoracic procedures has eliminated the concern about reentering the same operated side of the chest during spinal surgery. In some severe forms of scoliosis, an anterior release to the spine with multiple disc excisions, followed by a posterior fusion result in a higher degree of curve correction than a posterior spinal fusion alone. Although our patient was 13 years of age, she was skeletally immature, and a posterior spinal arthrodesis alone would likely have resulted in a crankshaft phenomenon and an increasing scoliosis, despite a solid posterior fusion. Under these circumstances, a first-stage anterior spinal fusion was required to control the deformity. Our case report shows that elective anterior and posterior spinal arthrodesis can be safely performed in a cardiac transplant recipient with severe developmental scoliosis, and can achieve optimum correction of the curvature and a successful out-

come without long-term medical or technical complications.

■ Key Points

- Cardiac transplantation has increased life expectancy in children with severe forms of congenital heart disease.
- The association of scoliosis with congenital cardiac disease is well established.
- To our knowledge, there is only 1 report of a patient with cardiac transplant and developmental scoliosis treated surgically, and this was performed with a posterior-only spinal arthrodesis.¹⁸
- Anterior scoliosis surgery has been previously contraindicated after heart transplantation.
- This case report has shown that elective anterior and posterior spinal arthrodesis can be successfully performed in cardiac transplant recipients with severe developmental scoliosis.

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Spinal Deformity in Patients With Sotos Syndrome (Cerebral Gigantism)

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and Michael J. McMaster, MD, FRCS

Study Design: Retrospective review of a case series.

Objective: To present the clinical characteristics and progression of spinal deformity in patients with Sotos syndrome.

Summary of Background Data: There is limited information on the development of spinal deformity and the need for treatment in this condition.

Methods: The medical records and spinal radiographs of 5 consecutive patients were reviewed. All patients were followed to skeletal maturity (mean follow-up: 6.6 y).

Results: The mean age at diagnosis of spinal deformity was 11.9 years (range: 5.8 to 14.5) with 4 patients presenting in adolescence. The type of deformity was not uniform. Two patients presented in adolescence with relatively small and nonprogressive thoracolumbar and lumbar scoliosis, which required observation but no treatment until the end of spinal growth. Three patients underwent spinal deformity correction at a mean age of 11.7 years (range: 6 to 15.4). The first patient developed a double structural thoracic and lumbar scoliosis and underwent a posterior spinal arthrodesis extending from T3 to L4. Five years later, she developed marked degenerative changes at the L4/L5 level causing symptomatic bilateral lateral recess stenosis and affecting the L5 nerve roots. She underwent spinal decompression at L4/L5 and L5/S1 levels followed by extension of the fusion to the sacrum. The second patient developed a severe thoracic kyphosis and underwent a posterior spinal arthrodesis. The remaining patient presented at the age of 5.9 years with a severe thoracic kyphoscoliosis and underwent a 2-stage antero-posterior spinal arthrodesis.

Conclusions: The development of spinal deformity is a common finding in children with Sotos syndrome and in our series it occurred in adolescence in 4 out of 5 patients. There is significant variability on the pattern of spine deformity, ranging

from a scoliosis through kyphoscoliosis to a pure kyphosis, and also the age at presentation and need for treatment.

Key Words: Sotos syndrome, cerebral gigantism, scoliosis, spinal deformity, treatment

(*J Spinal Disord Tech* 2009;22:149–153)

In 1964, Sotos et al¹ described 5 children with a condition characterized by a static cerebral disorder, excessively rapid growth in infancy, acromegalic features, a high-arched palate, and prominent jaw. They suggested that the triad of prenatal and postnatal overgrowth, learning difficulties, and a typical facial appearance constitutes a distinct clinical entity.¹ The condition was initially called “cerebral gigantism” because of the characteristic head shape and size, and the fact that it was thought to be due to a discrete lesion in the brain related to the hypothalamus. In a subsequent series of 6 patients, reported by Hook and Reynolds,² extensive endocrinologic investigations showed no abnormalities of the hypothalamic-pituitary axis and the term cerebral gigantism was abandoned as misleading in favor of the eponymous Sotos syndrome.

Since these 2 initial reports, a number of Sotos syndrome series were published which proposed a number of associated clinical features (Table 1).^{3–12} However, the diagnostic criteria for the condition were only established in 1994 by Cole and Hughes,¹³ who systematically assessed the clinical features and facial photographs of 79 patients with a provisional diagnosis of Sotos syndrome and evaluated their photographs between ages 1 and 6 years. They identified 41 probands that they considered to have the classic Sotos gestalt, compared their phenotype including anthropometric measurements, bone age, presence of learning difficulties, and associated medical comorbidities with the remaining 38 probands, and found significant differences between the 2 groups. They then proposed that Sotos syndrome was a distinct clinical syndrome characterized by 4 major diagnostic criteria: overgrowth, characteristic facial appearance, advanced bone age, and learning difficulties.¹³

Kurotaki et al¹⁴ were the first to identify a link between the clinical phenotype of Sotos syndrome and the mutation in the *NSD1* gene (nuclear receptor-binding SET domain containing protein). Mutations and micro-deletions of the *NSD1* gene on chromosome 5q35 have

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TABLE 1. Clinical Features in Sotos Syndrome (cerebral gigantism)¹⁻¹²

Facial Features	Musculoskeletal	Cognitive/Behavioral	Miscellaneous/Systemic
Macrocephaly	Excessive initial overgrowth/adult height is usually normal	Nonprogressive learning difficulties, attention deficit disorder, and behavioral problems (including phobias, aggression, obsessions, adherence to routine) in varying degrees from the individual living an independent life to being completely dependent on home care	Genitourinary anomalies
Long, thin face with prominent forehead, narrow skull, fronto-temporal hair sparsity, and flat-bridged nose	Acromegalic features Pes planus Generalized ligamentous laxity with hyperextensible joints		Tumors Paraspinal neuroblastoma Ganglioglioma Sacrococcygeal teratomas Presacral ganglioneuroma Gastric carcinoma Small-cell lung cancer Wilms' tumor Acute lymphocytic leukemia Congenital heart anomalies Neonatal jaundice/ hyperthyroidism/ hypothyroidism Seizures/nystagmus/strabismus Constipation/megacolon
Ruddy cheeks Down slanting palpebral fissures	Congenital hypotonia Spinal deformity (most commonly scoliosis)		
Square or pointed chin High-arched palate with early eruption of teeth	Cervical instability	Autistic spectrum disorder	

been recognized as the cause of the condition.¹² No correlation between mutation/deletion size and phenotype has been identified.¹⁵ Most cases are sporadic, even though there have been reports of autosomal dominant transmission.¹⁶

The orthopedic manifestations of Sotos syndrome include scoliosis, cervical instability, pes planus, hyperextensible joints, which may all be secondary to generalized ligamentous laxity, and increased growth rates with the affected children being tall for their peer group but having a height that is usually in the upper limits of normal by adulthood.^{12,17-19}

The purpose of this report is to present 5 patients with Sotos syndrome who have developed spinal deformity and discuss the course of the deformity and the need for treatment up to skeletal maturity.

MATERIALS AND METHODS

We reviewed the medical notes and spinal radiographs of 5 consecutive patients (3 females and 2 males) with Sotos syndrome who were followed by the senior author between 1980 and 2007 in a tertiary spinal center. These patients were diagnosed with Sotos syndrome between the ages of 7 months and 4 years by pediatricians and medical geneticists on the basis of the typical clinical findings associated with the condition. None of the patients had a family history of Sotos syndrome. We did not have access on the whole group of patients with

Sotos syndrome, therefore, we cannot comment on the prevalence of scoliosis in this population within the same chronologic period.

The mean age at initial diagnosis of spinal deformity was 11.9 years (range: 5.8 to 14.5). The mean age at presentation in our spinal deformity clinic was 13.3 years (range: 5.9 to 16.9), with 4 of these patients presenting after their adolescent growth spur with a Risser grade of 3 or 4. All 5 patients were followed clinically and radiologically for their spine deformity to skeletal maturity (Risser grade 5).

The type of spinal deformity and curve pattern was assessed on anteroposterior and lateral radiographs of the spine made with the patient standing. The site of the curvature was defined according to the classification proposed by the Scoliosis Research Society; cervicothoracic (apex at C7 or T1), thoracic (apex between T2 and T11), thoracolumbar (apex at T12 or L1), lumbar (apex between L2 and L4), and lumbosacral (apex at L5 or caudad).²⁰ The serial spinal radiographs were measured in both the coronal and sagittal planes with the use of the Cobb method.²¹ Care was taken to measure all curves with use of the same anatomic landmarks. Radiographs of the cervical spine were also available in all patients.

Three patients underwent spinal deformity correction at a mean age of 11.7 years (range: 6 to 15.4). The mean postoperative follow-up was 8.3 years (range: 4 to 12).

RESULTS

The type of spinal deformity and course of treatment in our patient population are shown in Table 2. Despite an early diagnosis of Sotos syndrome in all our patients, the spinal deformity was not diagnosed until adolescence in 4 of these patients. None of our patients underwent brace treatment.

The patterns of spinal deformity were very variable. Two patients presented in their adolescent life with relatively small and nonprogressive scoliosis, which required observation but no treatment until the end of spinal growth. One of these patients developed a thoracolumbar scoliosis (patient 1, Table 2) and the other patient developed a lumbar scoliosis (patient 2, Table 2).

Two patients presented in adolescence with severe deformities requiring surgical treatment. One of these patients developed a double structural thoracic and lumbar scoliosis and underwent a posterior spinal arthrodesis extending from T3 to L4 with AO Universal Spinal System II (AO USS II) instrumentation and iliac crest autograft with a good outcome (patient 3, Table 2). However, 5 years later, the patient developed marked degenerative changes at the L4/L5 level causing symptomatic bilateral lateral recess stenosis and affecting the L5 nerve roots. These findings were confirmed with magnetic resonance imaging. She underwent spinal decompression at the L4/L5 and L5/S1 levels followed by extension of the instrumented arthrodesis to the sacrum. At the latest follow-up, 4 years after revision spinal surgery, the patient was free of symptoms and had a solid fusion with no loss of correction. The second patient developed a severe thoracic kyphosis and underwent a posterior spinal arthrodesis with AO USS II instrumentation and iliac crest autograft with a good outcome (patient 4, Table 2).

The remaining patient presented at the age of 5.9 years with an extremely severe thoracic kyphoscoliosis (patient 5, Table 2). She underwent a 2-stage anterior and posterior spinal arthrodesis with Luque instrumentation and rib autograft with a good outcome.

Cervical radiographs were obtained in all 5 patients and did not show evidence of instability.

DISCUSSION

Sotos syndrome is a genetic condition causing physical overgrowth during the first years of life. This rapid physical development is often accompanied by delayed motor, cognitive, and social development. Muscle tone is low, and speech is markedly impaired. Prolonged drooling and mouth breathing may be present because of poor tone of facial muscles. Delays in gross and fine motor development are marked in early childhood and improve during the school years. Receptive language tends to advance sooner than expressive language, setting the stage for frustration. Older children seem to develop competence and normal speech patterns. Borderline to average intelligence is expected, as well as learning deficits and poor development of social skills. Intellectual, social,

TABLE 2. Description of Spinal Deformities and Course of Treatment in Our Patient Population

Pt/Sex	Age Related to the Spinal Deformity (y)		Type of Spinal Deformity	Cobb Angle at Surgery (Latest Postop Follow-up) or Discharge (degrees)		Follow-up (y)	Management
	Diagnosis	Surgical Treatment					
1/F	14.5	N/A	Left thoracolumbar scoliosis T11-L3	37	4	4	Observation to skeletal maturity
2/M	14	N/A	Left lumbar scoliosis T12-L4	45	4	4	Observation to skeletal maturity
4/F	13.1	13.6	Right thoracic scoliosis T6-L2, Left lumbar scoliosis L2-L4	53 (corrected to 39), 67 (corrected to 40)	9	9	Posterior spinal fusion T3-L4 with AO USS II instrumentation/ lumbar decompression L4-S1 and extension fusion to the sacrum
3/M	12	15.1	Thoracic kyphosis T2-L1	97 (corrected to 47)	4	4	Posterior spinal fusion T4-L2 with AO USS II instrumentation
5/F	5.8	5.9	Right thoracic kyphoscoliosis T2-T10	Scoliosis: 111 (corrected to 27) kyphosis: 134 (corrected to 65)	12	12	Two-stage anterior and posterior spinal fusion T2-L4 with Luque instrumentation

N/A indicates non-applicable; postop, postoperative; Pt, patient.

and emotional maturity may evolve on widely different timetables.

Children with Sotos syndrome are often taller, heavier, and have larger heads than their peers. A child who looks older and acts younger than his/her peer group is at risk for poor self-esteem, strained peer and family relationships, and problems in school. Fortunately, in late childhood the gap begins to close. Muscle tone improves steadily and with it comes better speech. For many individuals, Sotos syndrome simply alters developmental timing; despite early trends, the adult with the condition is likely to be within the normal range of height and intellect. Coordination problems may, however, persist into adulthood.

There is limited information on the development of spinal deformity in patients with Sotos syndrome. Sotos et al¹ reported 1 patient with a mild scoliosis in their original series of 5 patients. Since this initial report, 3 additional patients have been described but the patients' clinical presentation and course of progression have not been detailed.^{18,22,23} Sweeney et al²⁴ presented 2 patients who developed a scoliosis at the age of 8 weeks and 10 months and were treated with bracing and surgical correction. Cole and Hughes¹³ reported 3 of 41 patients with Sotos syndrome who were found to have a kyphoscoliosis but none required treatment.

Scoliosis was also diagnosed in 43% (43/101) of the patients reported by Tatton-Brown and Rahman,¹² many of whom required either bracing or surgical correction. The type of spinal deformity has not been described. After this study, scoliosis has been recognized to be one of the commonest clinical associations of Sotos syndrome.

In 1996, Haga et al¹⁷ presented 5 patients with Sotos syndrome who developed a scoliosis between the ages of 6 months and 5 years and outlined the differences between the scoliosis observed in this condition and infantile idiopathic scoliosis. The progression of scoliosis was rapid by 4 years of age, which corresponds to the characteristic period of overgrowth in this syndrome. Despite brace treatment, the scoliosis also progressed rapidly after 8 years of age. It was suggested that the progression of scoliosis might be related to the effect of growth hormone. Surgical correction was performed in 3 patients at a mean age of 11.7 years. The authors recommended early bracing, close observation for brace compliance and curve progression, and surgery near skeletal maturity.¹⁷

In contrast, 4 of the 5 patients presented in our study were diagnosed with a spinal deformity during adolescence even though they were closely monitored since early childhood. There was no sex predominance. We noted variable patterns of spinal deformity with 2 patients having a thoracolumbar or lumbar scoliosis, and the remaining 3 patients developing a double structural scoliosis, a kyphoscoliosis, and a pure kyphosis. Brace treatment was not applied in any of our patients and the 2 patients with a thoracolumbar or lumbar scoliosis were followed to skeletal maturity with no progression of the deformity. The remaining 3 patients underwent surgical

treatment at a mean age of 11.7 years with a satisfactory outcome and no loss of deformity correction at the latest follow-up.

Carlo and Dormans¹⁹ reported a 3-year 4-month-old child with Sotos syndrome who developed a 50% anterolisthesis at C3/C4 causing neurologic compromise and requiring segmental posterior stabilization and fusion. None of the patients included in our series has developed cervical instability or neurologic complications before or after treatment.

In conclusion, Sotos syndrome is a rare overgrowth condition characterized by a unique constellation of symptoms and signs including macrosomia, facial dysmorphism, advanced bone age, learning difficulties, and spinal deformity. Ligamentous laxity and rapid skeletal growth may be factors that predispose patients with Sotos syndrome to develop a spinal deformity. There is significant variability on the pattern of this deformity, ranging from a scoliosis through kyphoscoliosis to a pure kyphosis, the age at presentation, and the need for treatment. Patients with this condition should be closely monitored from infancy to skeletal maturity for the development of spine deformity, so that early orthopedic referral can be made and where necessary, appropriate treatment can be commenced.

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Spinal Deformity in Patients With DiGeorge Syndrome

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and Michael J. McMaster, MD, FRCS

Study Design: Retrospective review of a case series.

Objective: To present the clinical characteristics and treatment of spinal deformity in DiGeorge syndrome.

Summary of Background Data: There are no data on the development of spinal deformity in this condition. A high rate of wound infection could be expected after spinal surgery due to congenital thymic hypoplasia.

Methods: The medical records and spinal radiographs of 4 consecutive patients were reviewed. All patients were followed for a minimum of 2.5 years after spinal surgery.

Results: The mean age at diagnosis of spinal deformity was 9.9 years. The type of deformity was not uniform but the curves progressed in all patients requiring a spinal arthrodesis at a mean age of 16.1 years. Two patients developed a thoracolumbar scoliosis and underwent an anterior spinal arthrodesis with instrumentation, which produced a good outcome. The remaining 2 patients developed a double thoracic and lumbar scoliosis with increased thoracic kyphosis. Brace treatment was unsuccessful to control the deformity in one of these patients. Both patients underwent a posterior spinal arthrodesis with segmental instrumentation, which achieved satisfactory correction of the scoliosis with no loss of correction or detected pseudarthrosis at follow-up. However, both patients developed junctional kyphosis above the proximal end of the instrumentation. This was asymptomatic and nonprogressive; therefore, cephalad extension of the fusion was not required. Irradiated blood products were administered in all patients to prevent graft-versus-host disease. The postoperative course was uncomplicated and none of the patients developed wound healing problems or infections.

Conclusions: The development of spinal deformity in patients with DiGeorge syndrome may be associated to the presence of marked ligamentous laxity and congenital cardiac disease. There is significant variability on the pattern of spinal deformity,

which in our series was progressive in all patients and required surgical correction with no perioperative complications.

Key Words: DiGeorge syndrome, microdeletion 22q11.2, scoliosis, spinal deformity, treatment

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DiGeorge syndrome is a condition characterized by hypoplasia of the parathyroid glands and the thymus and leading to neonatal hypocalcemia presenting as tetany or seizures, and immunodeficiency due to decreased T-cell numbers with variable functional deficits.^{1–6} The phenotypic expression of the condition is due to disturbance of cervical neural crest migration affecting the derivatives of the pharyngeal arches and causing a defective embryologic development of the third and fourth pharyngeal pouches.

The condition is caused by a de novo 1.5 to 3 megabase hemizygous deletion in chromosome 22q11.2 mediated by low copy number repeats.^{7–9} Deletion of 22q11.2 occurs in about 1 in 4 to 6,000 newborns and is associated with a spectrum of clinical phenotypes including DiGeorge syndrome, as well as conotruncal anomaly facial syndrome and Shprintzen or velocardiofacial (VCF) syndrome.^{10–13}

Scambler et al¹⁴ first demonstrated microdeletions of chromosome 22q11 in patients with DiGeorge syndrome using fluorescent in situ hybridization (FISH). Kelly et al¹⁵ found similar genetic abnormalities in 9 patients with VCF syndrome. Driscoll et al¹⁶ confirmed the common genetic background of abnormality between DiGeorge and VCF syndrome with 90% of patients with DiGeorge and 75% of those with VCF having an interstitial deletion of 22q11. This evidence led Wilson et al¹⁷ to propose the term CATCH22 (Cardiac abnormality, Abnormal facies, T cell deficit due to thymic hypoplasia, Cleft palate, and Hypocalcemia due to hypoparathyroidism) as a collective acronym for those conditions with a common genetic etiology.

The diagnosis of DiGeorge syndrome is usually based on the presence of a dysmorphic facial appearance associated with outflow tract defects of the heart, and a history of recurrent infections. Hypocalcemia is the predominant biochemical feature and this may be symptomatic, even though there is a tendency for spontaneous resolution in early childhood. Further assessment includes a chest radiograph to detect a thymic shadow, and measurement of the CD4-positive subset of

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white cells, which demonstrates deficiency of T cells. With recent developments in molecular cytogenetics, genetic confirmation of the diagnosis investigating the karyotype and using FISH should be performed and the parents should be screened for carrier status.

The clinical features of DiGeorge syndrome include cardiac malformations affecting the outflow tract and these involve tetralogy of Fallot, and vascular dysgenesis with defects in the great vessels (Table 1¹⁻²³). Other associated abnormalities include facial dysmorphism with hypertelorism, telecanthus with short palpebral fissures, upward or downward slanting eyes, mandibular hypoplasia, low-set ears with anomalous folding of the pinna and hearing deficits, hypernasal speech, and renal abnormalities and uterine/vaginal agenesis. In addition, central nervous system anomalies, intellectual disabilities with mild-to-moderate learning difficulties, and developmental delay are often present.

Medical management of patients with DiGeorge syndrome includes calcium supplements and 1,25-cholecalciferol to treat hypocalcemia. Cardiac assessment and correction of heart or vascular defects is the primary focus of treatment in childhood. Early thymus transplantation may promote immune recovery if performed before the development of recurrent infections. Any affected child with confirmed immunodeficiency undergoing major surgery should have a supply of irradiated blood to avoid graft-versus-host disease. Educational support and speech therapy may also be required.

To the authors' knowledge there is no previous study to investigate the development of spinal deformity in patients with DiGeorge syndrome. The purpose of this report is to present 4 patients with DiGeorge syndrome who have developed spinal deformity and discuss the pattern and course of the deformity, and the need for treatment. Our hypothesis is that due to the congenital hypoplasia of the thymus and the inherent immunodeficiency these patients would be expected to carry a high risk for wound infection after spinal surgery.

MATERIALS AND METHODS

We reviewed the medical notes and spinal radiographs of 4 consecutive patients (2 males and 2 females) with DiGeorge syndrome. All patients were diagnosed

with a spinal deformity by their pediatricians who referred them to our clinic for further assessment and treatment. These patients were diagnosed with DiGeorge syndrome in early childhood by pediatricians and medical geneticists based on the typical clinical features associated with the condition. Genetic diagnosis using FISH was obtained in all patients and demonstrated deletions at chromosome 22q11.2. None of the patients had a family history of DiGeorge syndrome and the parents were not found to be carriers. None of our patients had affected siblings with the condition. We did not have access to the whole population of patients with DiGeorge in our area, therefore, we cannot comment on the overall prevalence of scoliosis in this condition.

The mean age at initial diagnosis of spinal deformity was 9.9 years (range: 3 to 14.7). The mean age at referral to our spinal deformity clinic was 12.7 (range: 3.3 to 16.9) with 3 of our patients presenting after their adolescent growth spur with a Risser grade of 2 to 4 (patients 1, 2, and 4, Table 2).

The type of spinal deformity and curve pattern were assessed on posteroanterior and lateral radiographs of the spine with the patient standing. The site of the curvature was defined according to the classification system proposed by the Scoliosis Research Society: cervicothoracic (apex at C7 or T1), thoracic (apex between T2 and T11), thoracolumbar (apex at T12 or L1), lumbar (apex between L2 and L4), and lumbosacral (apex at L5 or caudad).²⁴ The serial spinal radiographs were measured in both coronal and sagittal planes with the use of the Cobb method. Care was taken to measure all curves using the same anatomic landmarks. None of our patients had congenital vertebral anomalies.

All 4 patients underwent spinal deformity correction at a mean age of 16.1 years (range: 15.9 to 16.9). Our patients were followed clinically and radiographically for a minimum of 2.5 years postoperatively (range: 2.5 to 5.5). At the latest follow-up all patients apart from patient 3 had reached skeletal maturity with a Risser grade of 5.

Other Skeletal Problems

Two patients had short stature. In addition, 2 patients had pectus excavatum. Low muscle tone, marked ligamentous laxity with positive wrist and Steinberg

TABLE 1. Clinical Features in DiGeorge Syndrome¹⁻²³

Facial Features	Cardiac Malformations	Musculoskeletal	Miscellaneous/Systemic
Hypertelorism	Tetralogy of Fallot	Scoliosis	Hypoparathyroidism/hypocalcemia
Telecanthus with slanting eyes	Interrupted aortic arch	Hypotonia	Thymic hypoplasia/deficiency of T cells
Low set ears with hearing deficits	Truncus arteriosus with pulmonary stenosis	Abnormal joint mobility (ranging from hypermobility to flexion contractures)	Infections/autoimmune disease
Bulbous nose with square nasal tip	Right aortic arch	Talipes equinovarus/metatarsus adductus	Renal abnormalities/uterine or vaginal agenesis/hypothyroidism
Hypernasal speech	Aberrant subclavian artery	Epiphyseal dysplasia	Intellectual/learning disabilities/schizophrenia
Mandibular hypoplasia/micrognathia		Short stature/pectus excavatum	Developmental delay/obesity/cholelithiasis
Cleft palate and lip			

thumb sign combined with slender fingers, and mild camptodactyly was noted in all 4 patients. None of our patients had cervical instability. A magnetic resonance imaging of the spine was performed despite normal neurologic findings on clinical examination due to the syndromic condition and this was normal in all patients. There was no patient with meningocele. None of the 4 patients was affected by juvenile rheumatoid arthritis or other autoimmune disease.

Associated Systemic Anomalies

Two of our patients had congenital cardiac abnormalities, who were treated successfully in early childhood; these included tetralogy of Fallot and ventriculoseptal defect with infundibular pulmonary stenosis. An absent and a hypoplastic thymus gland were observed intraoperatively in the 2 patients who underwent cardiac surgery. Two patients had a history of recurrent chest infections in childhood, which settled as they reached puberty. Three patients had significant learning difficulties and required regular support at school. Two patients

had speech problems due to velopharyngeal insufficiency and 2 had hearing deficits. Two patients underwent cleft palate and 1 patient umbilical hernia repair. One patient had epilepsy. All patients had delayed motor and cognitive development. Renal function was normal in all patients.

RESULTS

The type of spinal deformity and course of treatment in our patient population are shown in Table 2. Despite an early diagnosis of DiGeorge syndrome in all our patients, the spinal deformity was not diagnosed until adolescence in 3 of these patients.

Spinal Deformity

The patterns of deformity were variable. Two of our patients were referred in adolescent life with moderate-to-severe thoracolumbar scoliosis that was relatively flexible (patients 1 and 2, Table 2). No previous treatment was applied. They underwent an anterior spinal arthrodesis extending across the thoracolumbar and upper lumbar

TABLE 2. Description of Spinal Deformities and Course of Treatment in our Patient Population

Patient/ Sex	Age Related to the Spinal Deformity (y)		Type of Spinal Deformity (Risser Grade)	Cobb Angle at Surgery/ Maximum Side-bending/Postop and Follow-up	Postop Follow-up (y)	Management
	Diagnosis	Surgical Treatment				
1/F	8	16.9	Left thoracolumbar scoliosis T11-L4 (Risser 4)	Preop: 77 degrees Side bending: 45 degrees Postop: 25 degrees Follow-up: 25 degrees	5.5	Anterior spinal fusion T11-L4 with instrumentation and rib autograft
2/F	14.7	15.9	Left thoracolumbar scoliosis T11-L4 (Risser 3)	Preop: 40 degrees Side bending: 25 degrees Postop: 6 degrees Follow-up: 6 degrees	2.5	Anterior spinal fusion T11-L3 with instrumentation and rib autograft
3/M	3	15.9	Left thoracic scoliosis T4-T12 Right lumbar scoliosis T12-L4 Thoracic hyperkyphosis (Risser 0)	Preop: 80 degrees Side bending: 50 degrees Postop: 21 degrees Follow-up: 21 degrees Preop: 52 degrees Side bending: 25 degrees Postop: 20 degrees Follow-up: 20 degrees Preop: 64 degrees Lateral bolster hyperextension: 55 degrees Postop: 50 degrees Follow-up: 50 degrees	2.5	Initial brace treatment (age: 9-15.9 y) Posterior spinal fusion T3-L4 with segmental pedicle screw/ rod instrumentation and iliac crest autograft
4/M	14	15.9	Right thoracic scoliosis T4-T10 Left lumbar scoliosis T10-L3 Thoracic hyperkyphosis (Risser 3)	Preop: 51 degrees Side bending: 45 degrees Postop: 39 degrees Follow-up: 39 degrees Preop: 38 degrees Side bending: 25 degrees Postop: 18 degrees Follow-up: 18 degrees Preop: 88 degrees Lateral bolster hyperextension: 75 degrees Postop: 60 degrees Follow-up: 60 degrees	2.5	Posterior spinal fusion T2-L3 with segmental pedicle screw/ rod instrumentation supplemented by proximal sublaminar wires, iliac crest autograft, and allograft bone

F indicates female; M, male; postop, postoperative; preop, preoperative.

spine with transvertebral screw/rod instrumentation and rib autograft and had a good outcome.

The third patient (patient 3, Table 2) presented to our clinic at the age of 3.3 years with a 15-degree thoracic and a 20-degree lumbar scoliosis. He was followed closely and subsequently required brace treatment at the age of 9 years when his thoracic scoliosis progressed to 32 degrees whereas the lumbar curvature remained stable. He had no increased thoracic kyphosis at that stage. Despite bracing his scoliosis continued to progress and he also developed an increased proximal thoracic kyphosis. Surgical correction was delayed due to patient and parent refusal until the thoracic scoliosis gradually deteriorated to 80 degrees and the lumbar scoliosis to 52 degrees. The thoracic scoliosis and associated hyperkyphosis was only moderately flexible preoperatively (Table 2). The patient underwent a posterior spinal arthrodesis with segmental pedicle screw/rod instrumentation and iliac crest autograft at the age of 15.9 years with a good outcome (Fig. 1). Attention was taken during placement of the T3 pedicle screws to preserve the proximal facet joints to the instrumentation intact to prevent add-on kyphosis. However, at 6 months postsurgery, the patient was noted to develop a junctional kyphosis above the cephalad end of the implant measuring 15 degrees between the end-plate of the most proximally instrumented vertebra and the upper end-plate of the vertebra above, which did not progress at 2.5 years of follow-up and did not cause him any symptoms. There was also no loss of scoliosis correction across the thoracic and lumbar curves at latest follow-up.

The remaining patient (patient 4, Table 2) presented at age 15.1 years with a thoracic and lumbar scoliosis and increased upper thoracic kyphosis. No treatment had been given before the referral to our clinic. The thoracic and lumbar scoliosis was relatively flexible in contrast to the thoracic hyperkyphosis that was rigid (Table 2). He underwent a posterior spinal arthrodesis with segmental pedicle screw/rod instrumentation supplemented by proximal sublaminar wires and the use of a combination of autologous iliac crest and allograft bone (Fig. 2). Extensive facetectomies, and segmental posterior closing wedge osteotomies were performed across the apex of the thoracic kyphosis to increase flexibility. Sublaminar wires were used in the thoracic spine at the proximal points of fixation to increase stability of the construct and reduce the risk of pull-out failure, which could have been higher if pedicle screws were applied in the presence of the hyperkyphosis. The posterior arthrodesis using bone graft was extended 1 level proximal to the instrumentation due to the use of sublaminar wires at the cephalad end of the construct, which require removal of the cranial interspinous and supraspinous ligaments and this could destabilize this area of the spine. An additional anterior spinal release was not considered to be necessary. Despite the presence of a small proximal junctional kyphosis of 10 degrees that did not deteriorate over time, the patient had a good outcome, no complaints of his spine, and no loss of scoliosis correction at the latest follow-up.

In the perioperative period all patients were administered irradiated blood products to avoid graft-versus-host disease. The postoperative course was uncomplicated

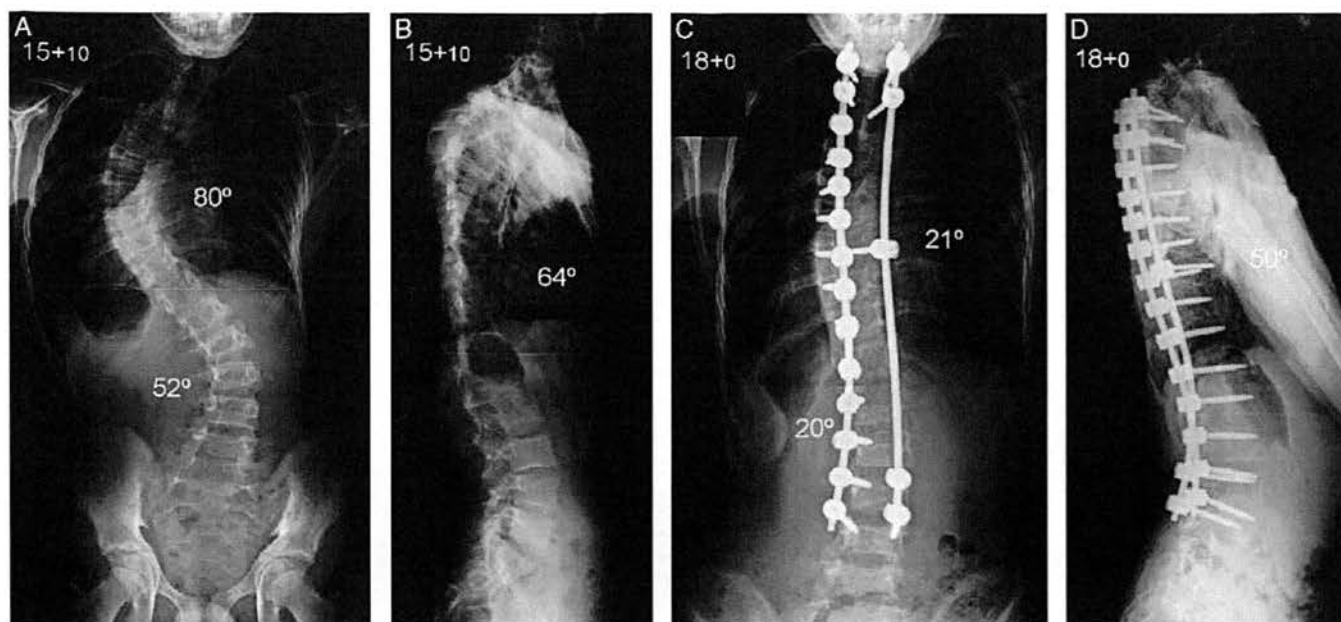


FIGURE 1. Preoperative posteroanterior (A) and lateral (B) radiographs of the spine of patient 3 show a left thoracic and right lumbar scoliosis with associated upper thoracic hyperkyphosis. Posteroanterior (C) and lateral (D) radiographs of the spine at 2.1 years after posterior spinal arthrodesis show a good correction of the scoliosis. There is evidence of a junctional kyphosis in the upper thoracic spine, measuring 15 degrees that was asymptomatic and did not progress.

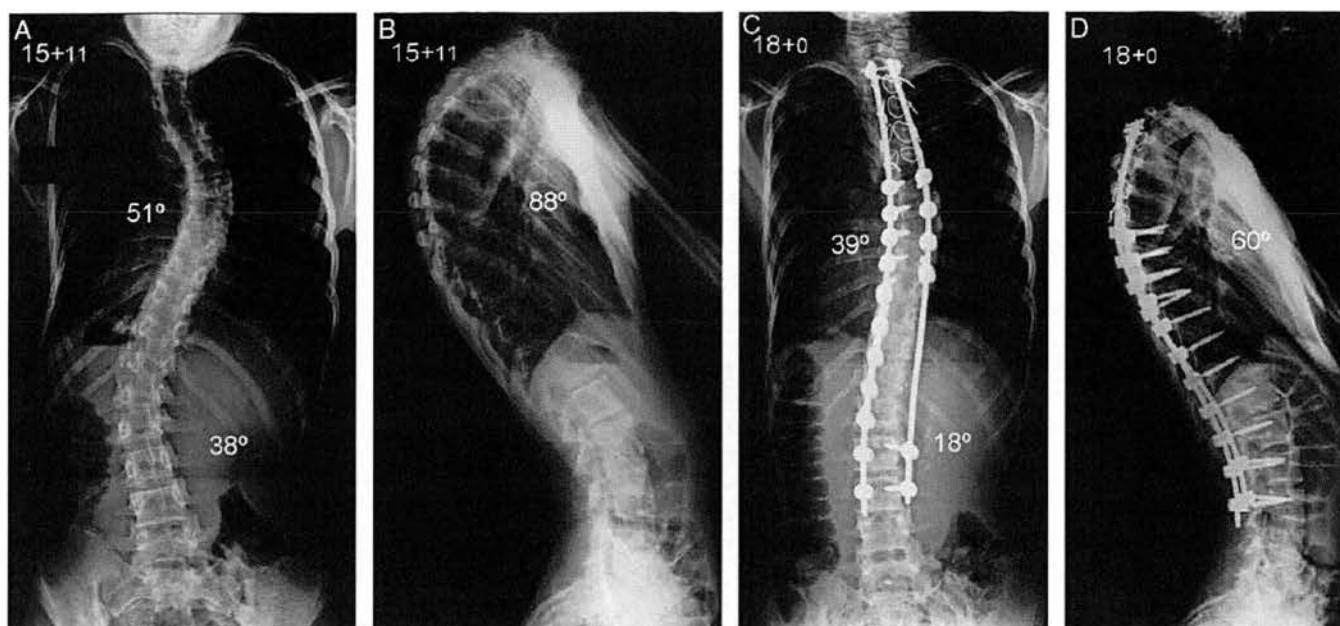


FIGURE 2. Preoperative posteroanterior (A) and lateral (B) radiographs of the spine of patient 4 show a right thoracic and left lumbar scoliosis with severe thoracic hyperkyphosis. Posteroanterior (C) and lateral (D) radiographs of the spine at 2.1 years after a posterior spinal arthrodesis show a good correction of the scoliosis and a small add-on kyphosis of 10 degrees in the upper thoracic spine.

in all 4 patients and none developed an infection or problems with wound healing. They were fitted with an underarm spinal brace to provide additional support to the spine and this remained for 4 months after surgery. In patients 1 and 2 who underwent an anterior spinal arthrodesis this followed the standard practice in our institution. Patients 3 and 4 were provided with a standard thoracolumbosacral orthosis due to the presence of thoracic hyperkyphosis preoperatively, to limit trunk flexion. The thoracolumbosacral orthosis postoperatively was not used to control high thoracic kyphosis. A Milwaukee brace would have been more effective in decreasing the risk of upper thoracic fixation pull-out and pseudarthrosis but this would have been very poorly tolerated by our patients. The mean hospital stay was 11 days (range: 9 to 12).

DISCUSSION

The skeletal manifestations of chromosome 22q11.2 deletion syndrome which incorporates the phenotype of DiGeorge syndrome include short stature, hypotonia, scoliosis, abnormal mobility of the joints ranging from flexion contractures affecting primarily the fingers to generalized ligamentous laxity, arachnodactyly of the fingers and toes, epiphyseal dysplasia, pectus excavatum, metatarsus adductus, and clubfoot.^{18–20} In our series, all 4 patients had low muscle tone with hypermobility of the joints and camptodactyly of the fingers; 2 patients had short stature; and 2 patients had pectus excavatum.

Scoliosis has been estimated to occur in approximately 10% of patients with chromosome 22q11.2

deletion syndrome.^{18,21–23} Morava et al¹⁹ evaluated 20 patients with VCF syndrome and described 3 children (15%) who developed a spinal deformity. Only one of these patients underwent an uneventful posterior spinal arthrodesis to correct a double thoracic and thoracolumbar scoliosis with associated thoracolumbar kyphosis. The remaining 2 patients presented with double thoracic and thoracolumbar scoliosis, which did not require treatment.

All 4 patients included in our study presented with progressive spinal deformities and necessitated surgical correction. We noted variable patterns of spinal deformity with 2 female patients having a thoracolumbar scoliosis, and the remaining 2 male patients developing a thoracic and lumbar scoliosis with associated thoracic hyperkyphosis. Even though the number of patients is too small to extract meaningful conclusions, it is interesting to note that the male and female patients in our series have developed completely different patterns of spinal deformity. Brace treatment was applied in only one of our male patients and this was ineffective to control the deformity. Surgical correction of the scoliosis or kyphoscoliosis was performed in all patients at a mean age of 16.1 years and this was not associated with any postoperative complications. A satisfactory surgical outcome was achieved and there was no loss of correction across the levels of the instrumented arthrodesis and no detected pseudarthrosis at the latest clinical and radiographic follow-up (mean: 3.25 y after surgery).

However, both male patients who underwent a posterior spinal arthrodesis for a thoracic and lumbar scoliosis associated with increased thoracic kyphosis

developed postoperatively an asymptomatic junctional kyphosis above the proximal level of the instrumented fusion. Patient 3 underwent a posterior spinal arthrodesis using a pedicle screw/rod construct with the proximal end of the rods contoured with no thoracic kyphosis. In patient 4, sublaminar wires were placed in the upper thoracic spine to secure fixation of the rods to the spine, which necessitate removal of the cranial interspinous and supraspinous ligaments and this could destabilize this area. These could both be factors contributing to the development of the junctional upper thoracic kyphosis in our patients. Preserving the proximal facet joints to the instrumentation intact in patient 3 and extending the arthrodesis using bone graft to 1 level cephalad to the implant in patient 4 has not prevented the add-on kyphosis in the upper thoracic spine. It is uncertain whether a proximally longer fusion would have reduced the risk of this add-on kyphosis while at the same time this could destabilize the cervicothoracic junction and cause further junctional problems. The junctional kyphosis was nonprogressive at follow-up to date with both our patients being skeletally mature; therefore, proximal extension of the fusion has not been required.

The association of scoliosis with congenital cardiac disease is well established.²⁵⁻²⁷ The majority of cases involve developmental curvatures, resembling to idiopathic scoliosis. There is usually no evidence of congenital abnormalities in the formation of the vertebral column. The incidence of scoliosis in patients with congenital heart disease has been reported to range between 4% in those patients who have not received operative treatment and 11% in children who have undergone corrective surgery.²⁵⁻²⁷ Two of our patients had severe congenital cardiac disease requiring surgical treatment at an early age and they had no congenital vertebral anomalies. In these patients the presence of congenital heart abnormalities could have been a contributing factor in the development of scoliosis.

Morava et al¹⁹ suggested that an associated connective tissue disorder in patients with VCF syndrome could be the cause of the increased frequency of scoliosis. Haploinsufficiency of the *TBX1* gene, which is located within the chromosome 22q11.2, is responsible for most of the physical malformations in patients with DiGeorge syndrome and these deletions of *TBX1* may play a role in the development of connective tissue disease in this condition. In addition, Chieffo et al²⁸ showed that the mouse homolog of this gene is expressed in the fetal pharyngeal arches and pouches, and later in development in the musculoskeletal tissue. All our patients had joint hypermobility with arachnodactyly and camptodactyly, and 1 patient had an umbilical hernia. These are clinical signs suggestive of connective tissue abnormality, which we believe could account at least partly for the development of spinal deformity in our group of patients. It is also possible that the presence of marked inherent ligamentous laxity was a contributing factor to the development of junctional kyphosis after spinal deformity correction above the proximal end of the instrumented

fusion in the 2 patients with increased preoperative thoracic kyphosis.

Clinical features related to immunodeficiency associated with deletion of 22q11.2 and DiGeorge syndrome include susceptibility to infection, juvenile rheumatoid arthritis, idiopathic thrombocytopenia purpura, and autoimmune hemolytic anemia.² These are more common in patients with the most disordered T-cell function. None of our 4 patients were affected by autoimmune disease or recurrent infections at the time of spinal deformity surgery.

In conclusion, ligamentous laxity and congenital cardiac disease may be factors that predispose patients with DiGeorge syndrome to develop spinal deformities. We recorded significant variability on the pattern of this deformity ranging from an isolated thoracolumbar scoliosis in 2 female patients to a thoracic and lumbar scoliosis with associated thoracic hyperkyphosis in the remaining 2 male patients. In our patients, the deformity has been progressive and bracing was tried in only 1 patient who presented early with poor results. All 4 patients required surgical correction of their scoliosis or kyphoscoliosis with a successful outcome and no perioperative complications. Patients with this condition should be closely monitored from early childhood to skeletal maturity for the development of spine deformity so that adequate referral to a spinal clinic can be made and if necessary appropriate treatment can be applied.

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Rates of muscle protein synthesis in paraspinal muscles: lateral disparity in children with idiopathic scoliosis

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SUMMARY

1. The rate of paraspinal (multifidus) muscle protein synthesis was measured bilaterally at the top, apex and bottom of the thoracic curve in nine children with an idiopathic scoliosis, using the stable-isotope-labelled amino acid L-[1-¹³C]leucine.

2. No significant difference was observed in rates of muscle protein synthesis between the two sides of the spine, at the levels of the first vertebrae in neutral alignment at the top and bottom of the curve. However, in every patient, at the apex of the spinal curve, synthesis was higher on the convexity than on the concavity (0.077 ± 0.04 %/h convex, 0.052 ± 0.02 %/h concave, means \pm SD, $P < 0.01$).

3. Muscle RNA activity (μ g of protein synthesized h^{-1} μg^{-1} of RNA) was lower at the curve apices on the concave than the convex side (0.019 ± 0.09 $\mu\text{g h}^{-1} \mu\text{g}^{-1}$ convex apex, 0.016 ± 0.06 $\mu\text{g h}^{-1} \mu\text{g}^{-1}$ concave apex, $P < 0.05$). Activities were similar on the two sides at the top and bottom of the curve.

4. Differences in muscle histology between the two sides were also observed only at the apex, with a lower type I fibre diameter (50.9 ± 8.5 μm convex, 38.3 ± 2.4 μm concave, $P < 0.05$) and a lesser proportion of type I fibres ($63 \pm 12\%$ convex, $49 \pm 9\%$ concave, $P < 0.05$) on the concavity.

5. The results are consistent with effects on muscle protein turnover secondary to an increased muscle contractile activity on the curve convexity and functional immobilization of the muscle on the curve concavity.

Key words: immobilization, protein synthesis, scoliosis, skeletal muscle.

Abbreviations: APE, atoms per cent excess; α -KIC, α -ketoisocaproate; GC-MS, gas chromatography-mass spectrometry; IRMS, isotope ratio mass spectrometry; k_e , muscle protein synthetic rate.

INTRODUCTION

In patients with idiopathic scoliosis, histological and histochemical abnormalities of the paraspinal muscles have been reported. Several investigations have confirmed a preponderance of type I muscle fibres in multifidus at the apex of the curve on the convex side [1-4], and electromyogram studies have also shown greater electrical activity on the curve convexity [5]. We reasoned that differences in muscle contractile activity in the presence of altered fibre type proportions would be accompanied by alteration in muscle protein turnover [6]. The only previous data are from measurements *in vitro* of protein turnover in muscle sampled from patients with scoliosis of mixed aetiological types [7]. These studies provided inconclusive evidence that muscle protein synthesis might be elevated on the curve convexity. Until recently, methods of measuring muscle protein synthesis *in vivo* could not be applied in growing children, because they involved the use of radioactive isotopes.

The purpose of this paper is to report the results of the first measurements of the rate of paraspinal muscle protein synthesis *in vivo* in children with idiopathic scoliosis, using stable-isotope technology [8].

MATERIALS AND METHODS

Nine children with an adolescent idiopathic right thoracic scoliosis requiring surgical correction and spine fusion, were studied at the Scoliosis Unit, Princess Margaret Rose Orthopaedic Hospital, Edinburgh. There were eight girls and one boy whose mean age was 14 years 6 months (range 12 years 4 months-17 years 9 months) and mean weight 48 kg (range 34-63 kg). Ethical approval was

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granted by the Lothian Health Board Research Ethics Committee and informed parental consent was given for each investigation.

Pulmonary function (vital capacity, forced expiratory volume and peak expiratory flow rate) was measured pre-operatively in each child using standard physiological methods [9]. Each child was fasted for the 12 h preceding spinal surgery. Eight hours before operation, L-[1-¹³C]-leucine (1 mg/kg) and NaH¹³CO₃ (0.16 mg/kg), both 99% ¹³C (Tracer Technologies Inc., Newton, MA, U.S.A.), were administered as an intravenous priming bolus [10]. This was followed by a continuous intravenous infusion of L-[1-¹³C]-leucine at a rate of 1 mg h⁻¹ kg⁻¹ [11] via a cannula in a forearm vein. Blood was sampled every 15 min for the first 2 h of infusion then hourly thereafter via a cannula placed in a vein in the contralateral forearm. The final sample was taken after induction of anaesthesia, 10 min before muscle sampling.

After an initial trial collection, two separate timed (7 min) collections of expired gas were made into 50 litre Douglas bags. Expired volumes were measured with a dry gas meter (Harvard Apparatus Ltd, Edenbridge, U.K.) and production of CO₂ was estimated from the CO₂ concentration (measured by i.r. absorbance gas analyser; Grubb Parsons Ltd, Newcastle, U.K.). Variation in expired CO₂ volume per minute between the two ventilations was <5%. Hourly samples of expired CO₂ were collected throughout the infusion period into evacuated glass tubes (Vacutainer, Slough, U.K.).

One and a half hours before surgery (07.30 hours) each patient received oral pre-medication (10 mg of diazepam) and an injectable cephalosporin antibiotic (250 mg of cephadrine). General anaesthesia was induced (09.00 hours) and was maintained with N₂O/O₂ and isoflurane inhalation. The infusion of labelled leucine was continued until surgical exposure of the paraspinal muscles had been completed (approximately 09.40 hours). Multifidus muscle biopsies of approximately 200 mg wet weight were taken from both sides of the spine at the top, apex and bottom of the scoliosis, i.e. at the levels of the upper neutral, apical and lower neutral vertebrae.

Sample analysis

The samples were analysed using identical methods to those previously reported [6]. From measurements of the plateau enrichment with ¹³C of α -ketoisocaproic acid (α -KIC) in plasma (measured by gas chromatography-mass spectrometry, GC-MS) and of CO₂ in expired breath (measured by isotope ratio mass spectrometry, IRMS), whole-body leucine flux (equivalent in the fasted state to whole-body protein breakdown) and its components leucine oxidation and whole-body protein synthesis were calculated [8, 12]. The coefficient of variation in enrichment with ¹³C between expired gas samples taken at an isotopic steady state of plasma [¹³C]leucine labelling was <5%. Muscle protein synthesis (k_p) was calculated by comparing the increase in ¹³C enrichment of leucine incorporated into muscle protein with that of the time averaged steady-state plasma ¹³C α -KIC enrichment

which was taken as an index of the amino acid precursor pool enrichment (measured by IRMS and GC-MS, respectively) [10, 11, 13, 14]. Post-infusion muscle enrichments were of the order of 0.01–0.03 atoms per cent excess (APE) above a basal [¹³C]leucine enrichment of muscle protein, derived from the mean enrichment of six multifidus biopsies taken from two age-matched non-infused patients with idiopathic scoliosis. This basal enrichment approximated to 0.005 ± 0.0005 APE (mean \pm SD) measured against our routine CO₂ gas standards. Duplicate analyses of enriched samples agreed within a precision of $\pm 2\%$ on 50 mg wet weight biopsy samples. Plateau plasma enrichment of α -KIC (within the range 2.5–6.0 APE) occurred within 30 min of commencing the constant leucine infusion. Standard deviation between samples at plateau was <0.3 APE. No reproducible change in plasma α -KIC enrichment occurred after either pre-medication or induction of anaesthesia. In any case, alteration of the enrichment of the intracellular leucine pool would have only a small effect on the measured rate of muscle protein synthesis, since the period of anaesthesia was never longer than 40 min.

Nucleic acids were extracted from muscle [15] and assayed by double-wavelength spectrophotometry [16]. Alkali-soluble protein concentration was determined by a standard method [17].

Transverse 10 μ m mounted cryostat sections of muscle from each of the six biopsy sites per patient were processed. Type I, IIa and IIb fibres were identified by the magnesium adenosine triphosphatase activity of actomyosin at pH 9.4 in the presence of 10 mmol/l CaCl₂ [18]. From photomicrograph prints of each adenosine triphosphatase-stained specimen, the mean lesser diameters of about 50 fibres of each type were determined using an Imagen semi-automatic image analyser (Graphics Information Systems, Blairgowrie, U.K.). Elliptical and distorted fibres were excluded from the measurements.

Statistical analysis

Results are expressed as means \pm SD. Values from the two sides of the curve at any level were compared by using the Student's *t*-test for paired data.

RESULTS

The severity of the mean lateral curvature of the spine (Cobb angle) [19] before surgery in the nine children was 47 degrees (range 33–62 degrees). Vital capacity, forced expired volume in 1 s and peak expiratory flow rate were $89 \pm 13\%$, $76 \pm 11\%$ and $80 \pm 17\%$ (means \pm SD) of their normal predicted values.

Muscle histology showed no significant differences in fibre diameter or percentage of the two fibre types between muscle from the two sides of the spine, at the levels of the upper and lower neutral vertebrae (Table 1). However, at the apex of the curve on the concave side, there were type I fibres with a mean fibre diameter 12 μ m smaller, and 14% fewer type I fibres ($P < 0.05$), than on the curve convexity. The increase in type I fibres at the

apex on the convex side was accompanied by a 50% fall in the ratio of type IIb to IIa fibres ($P < 0.05$).

The mean values for multifidus muscle protein synthesis are shown in Table 2. There was no significant difference in the rates of muscle protein synthesis between the two sides of the spine at the levels of either the upper or lower neutral vertebrae. However, muscle protein synthesis in all of the patients was significantly higher on the convex side at the apex of the curve, than on the concave side ($P < 0.01$; Table 3). Two-way analysis of

variance indicated a significant difference ($P < 0.025$) between the mean muscle protein synthesis values at the three levels of biopsy on the convex side but not between those on the concave side ($P > 0.05$). On the convexity, muscle protein synthesis was significantly greater at the curve apex than at the lower level ($P < 0.02$, Student's paired t -test), but the values at the apex did not significantly exceed those from the top of the curve ($P > 0.05$). On the concave side, muscle protein synthesis was lower at the apex than at the upper level only ($P < 0.05$). RNA

Table 1. Multifidus muscle morphometry

Diameter = mean lesser diameter. Values are means \pm SD ($n = 9$). Statistical significance: * $P < 0.05$ (Student's paired t -test).

	I		IIa		IIb	
	Diameter (μm)	Proportion (% of total fibres)	Diameter (μm)	Proportion (% of total fibres)	Diameter (μm)	Proportion (% of total fibres)
Convex top	49.7 \pm 10.1	51 \pm 6	44.1 \pm 9.4	31 \pm 3	48.7 \pm 12.0	18 \pm 10
Convex apex	50.9 \pm 8.5	63 \pm 12	43.8 \pm 4.8	26 \pm 8	43.5 \pm 1.2	11 \pm 5
Convex bottom	51.0 \pm 5.2	54 \pm 8	41.9 \pm 5.1	20 \pm 3	45.5 \pm 2.4	26 \pm 4
Concave top	56.4 \pm 9.8	47 \pm 8	49.6 \pm 6.2	31 \pm 9	52.1 \pm 6.2	22 \pm 8
Concave apex	38.3 \pm 2.4*	49 \pm 9*	47.0 \pm 4.2	25 \pm 2	40.0 \pm 5.9	26 \pm 7
Concave bottom	49.8 \pm 7.4	53 \pm 3	42.9 \pm 3.8	26 \pm 11	42.5 \pm 3.5	21 \pm 8

Table 2. Muscle protein synthesis (k_s) in idiopathic scoliosis

Values are means \pm SD ($n = 9$). Statistical significance: * $P < 0.05$, ** $P < 0.01$ (Student's paired t -test).

		[Protein] (% wet weight)	[RNA] ($\mu\text{g}/\text{mg}$ of protein)	[DNA] ($\mu\text{g}/\text{mg}$ of protein)	RNA/DNA	k_s (%/h)	k_s/RNA (μg of protein $\text{h}^{-1} \mu\text{g}^{-1}$ of RNA)
Top	Convex	19.4 \pm 1.6	3.8 \pm 0.4	1.8 \pm 0.4	2.1 \pm 0.7	0.0707 \pm 0.04	0.20 \pm 0.10
	Concave	17.8 \pm 2.6	3.9 \pm 1.1	1.8 \pm 0.5	2.4 \pm 0.9	0.0657 \pm 0.03	0.19 \pm 0.12
Apex	Convex	17.6 \pm 1.9	4.0 \pm 0.6	2.2 \pm 0.8	2.2 \pm 1.1	0.0773 \pm 0.04	0.19 \pm 0.09
	Concave	17.8 \pm 1.8	3.3 \pm 0.6*	2.3 \pm 1.1	1.6 \pm 0.5*	0.0519 \pm 0.02**	0.16 \pm 0.06*
Bottom	Convex	17.8 \pm 1.8	3.8 \pm 0.7	1.8 \pm 0.6	2.3 \pm 0.8	0.0597 \pm 0.03	0.15 \pm 0.06
	Concave	18.3 \pm 2.6	3.6 \pm 0.7	1.8 \pm 0.5	2.2 \pm 0.7	0.0628 \pm 0.04	0.18 \pm 0.13

Table 3. Multifidus protein synthesis at the curve apex

Values are means \pm SD. Statistical significance: * $P < 0.01$ (Student's paired t -test).

Patient no.	Age (years/months)	Weight (kg)	Curve (degrees)	Top		Apex		Bottom	
				Convex k_s (%/h)	Concave k_s (%/h)	Convex k_s (%/h)	Concave k_s (%/h)	Convex k_s (%/h)	Concave k_s (%/h)
1	15/11	56	33	0.0365	0.0380	0.0355	0.0289	0.0280	0.0376
2	17/9	63	36	0.0501	0.0487	0.0562	0.0274	0.0427	0.0193
3	13/0	34	39	0.0727	0.0456	0.0679	0.0508	0.0601	0.0624
4	12/4	34	45	0.0616	0.0663	0.0638	0.0458	0.0573	0.0644
5	15/10	62	48	0.0627	0.0620	0.0541	0.0472	0.0606	0.0513
6	15/7	46	50	0.0991	0.0744	0.1402	0.0645	0.0770	0.0644
7	13/8	52	51	0.1527	0.1534	0.1497	0.0900	0.1151	0.1598
8	12/5	36	55	0.0416	0.0487	0.0643	0.0582	0.0386	0.0506
9	14/8	56	62	0.0591	0.0542	0.0638	0.0545	0.0582	0.0559
Mean \pm SD	14/6	49 \pm 12	47 \pm 9	0.0707 \pm 0.04	0.0657 \pm 0.03	0.0773 \pm 0.04	0.0519 \pm 0.02*	0.0597 \pm 0.03	0.0628 \pm 0.04

concentration, RNA/DNA ratio and rate of protein synthesis per unit RNA (RNA activity) were significantly lower in muscle specimens from the concavity at the apex of the curve, when compared with those from the convexity and also those from both sides at the level of the neutral vertebrae.

Whole-body protein turnover (i.e. leucine flux) in the nine patients with scoliosis was 32% greater than in normal healthy adults ($n = 25$, 14 male, 11 female, mean age 57 ± 18 years; J. N. A. Gibson & M. J. Rennie, unpublished work; Table 4).

DISCUSSION

The results presented here show that there is a difference in the rate of multifidus muscle protein synthesis between the two sides of the spine at the apex of an idiopathic scoliosis. In our opinion, this is likely to be a secondary phenomenon occurring as a consequence of the developing spinal deformity rather than contributing to its primary aetiology, since similar values of muscle protein synthesis were observed at the levels of the upper and lower neutral vertebrae. There are no comparable data from normal children of similar ages with which to compare the rates of muscle protein synthesis reported here. The values of multifidus muscle protein synthesis were at least 20% higher than our previous values obtained from adult quadriceps muscle of similar type I/type II fibre ratio (0.04%/h) [6]. Although in the present study the muscle biopsies were taken after 40 min of anaesthesia, which could have influenced the rate of incorporation of [^{13}C]leucine into muscle protein, it is unlikely that errors in calculation of muscle protein synthetic rate could have amounted to as much as 20% when precursor pool enrichment (plasma ^{13}C -KIC) remained constant up to the time of biopsy. Differences in k_e attributable to sex distribution can also probably be discounted, since four of the children studied were pubertal. These comparative values therefore suggest that where muscle fibre proportions are similar, muscle protein turnover is higher in children than in adults. This conclusion is unsurprising given the background of results from animals of differing ages, but it is the first such demonstration in humans. The wide range in absolute k_e values observed in the children

with scoliosis probably reflects the differing developmental age of each patient, which in a small group studied *ad hoc* would be unlikely to exactly correlate with chronological age. Whole-body protein turnover was also greater in the children than adults. Since the values of leucine oxidation were similar in the two groups, it would appear that the major reason for the higher rate of flux presently observed was the 48% absolute increase in the amount of leucine used for protein synthesis.

In the presence of similar muscle protein concentration to that at other sites, there was a significantly lower RNA concentration at the apex of the curve on the concavity. This was presumably due to there being fewer ribosomes per cell rather than to an alteration in fibre number, since the RNA/DNA ratio was also lower at this site. In addition, the lower rate of protein synthesis per unit RNA would suggest a lower rate of transcription of DNA and/or a decrease in efficiency of translation of RNA [20].

In these studies we have observed a lower proportionate representation of type I fibres per unit muscle cross-sectional area on the concavity, and greater representation of type I fibres on the convexity of an idiopathic curve. These findings agree with those reported previously [2, 3, 21]. Ford *et al.* [4] reported that the differences in histology are not confined to the multifidus muscle but extend to the superficial (externally related) layer of erector spinae muscle. Previous studies have shown that after knee immobilization there is atrophy of type I fibres in quadriceps muscle associated with a diminution of quadriceps strength [22, 23]. In addition, in the presence of the type I fibre atrophy, a lower rate of quadriceps muscle protein synthesis was observed [6]. The very similar findings with regard to the muscle fibre types and rate of protein synthesis on the concavity of an idiopathic scoliosis, suggests that they may be associated with a relative diminution in muscle strength on the concave side compared with that on the convex. The increase in muscle protein synthesis on the convexity may occur as an adaptation to the increasing stretch associated with the deformity [24], whereas the decrease in muscle protein synthesis on the concavity may be due to the relative flaccidity and functional immobilization of the muscle.

Subsequent to demonstrations in dogs that unilateral electrical stimulation of the paraspinal muscle will induce

Table 4. Whole-body leucine turnover in children with idiopathic scoliosis, compared with 25 normal adults of mean age 57 (range 19–77) years (J. N. A. Gibson & M. J. Rennie, unpublished work)

Values are means \pm SD. Statistical significance: * $P < 0.01$ (Mann-Whitney U -test).

	Children with scoliosis ($n = 9$)	Normal adults ($n = 25$)
Leucine flux (\dot{Q}) ($\mu\text{mol h}^{-1} \text{kg}^{-1}$)	$179 \pm 47^*$	135 ± 30
Leucine oxidation (\dot{O}) ($\mu\text{mol h}^{-1} \text{kg}^{-1}$)	47 ± 22	46 ± 17
Leucine removed by synthesis (\dot{S}) ($\mu\text{mol h}^{-1} \text{kg}^{-1}$)	$132 \pm 45^*$	89 ± 22
Dietary intake (D) ($\mu\text{mol h}^{-1} \text{kg}^{-1}$)	0	0
Leucine derived from breakdown (\dot{B}) = ($\dot{Q} - \dot{D}$) ($\mu\text{mol h}^{-1} \text{kg}^{-1}$)	$179 \pm 47^*$	135 ± 30
\dot{O}/\dot{Q} (%)	27 ± 13	33 ± 8
\dot{S}/\dot{Q} (%)	73 ± 13	67 ± 8

inequalities of skeletal muscle force acting on the two sides of a spine, producing scoliosis [25], electrical stimulation of the muscle on the convexity of an idiopathic scoliosis has been used clinically to prevent progression of relatively small curves [26, 27]. Theoretically, the objective is to strengthen the muscle and so create a corrective or adaptive response of muscle growth. Our findings suggest that such an imbalance in multifidus muscle protein synthesis already exists, but the growth adaptation and strength of the muscle at the apex on the convexity is not sufficient to prevent curve deterioration. We have recently shown [28] that electrical stimulation of atrophic quadriceps muscle can induce an increase in muscle protein mass, probably by decreasing the rate of quadriceps protein breakdown. Thus it is possible that similar beneficial changes occur with appropriate paraspinal muscle stimulation in children with scoliosis.

In conclusion, we suggest that although the primary aetiology of idiopathic scoliosis remains unknown, differences in the rate of muscle protein synthesis and contractile strength occur secondarily in response to differences in muscle activity on the two sides of the spinal curve.

ACKNOWLEDGMENTS

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Section H

Ankylosing Spondylitis

Spinal Osteotomy in Ankylosing Spondylitis

Technique, Complications, and Long-Term Results

A series of 17 patients with ankylosing spondylitis who had lumbar spinal osteotomy have been reviewed. Fifteen of the patients have been followed up for an average of 10 years (2 died during the postoperative period) in an attempt to assess the long-term results of this procedure. We found that the degree of correction in the lumbar spinal region remained constant once the osteotomy had fully united but that deformity tended to recur in patients whose disease was still active in the thoracic and cervical regions, and this detracted from the early results of the operation. Hip flexion contracture also detracted from the result of spinal osteotomy but can be corrected by suitable hip surgery, during which the contracture is released. Significant complications can be expected from this procedure, but most patients are benefited both physically and psychologically.

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Ankylosing spondylitis, when allowed to run its full course without proper medical management, is one of the most crippling diseases seen by the orthopedic surgeon. The characteristic deformity is a flattening of the normal lumbar lordosis, with an exaggeration of the dorsal kyphosis from which the head and neck project forward in a straight line, or in flexion with the axis of the upper dorsal spinal column. Eventually, as the disease progresses, the entire spinal column from sacrum to occiput may become ankylosed by bone in this deformed position. At this stage, the patient is bent forward and forced to look at the ground. To see in front of him, he attempts to hyperextend his hips, but unfortunately this may not be possible if the hips are also involved (as they are in 42% of patients).¹ Therefore, the patient flexes his knees and so produces the characteristic stance seen in ankylosing spondylitis (Fig. 1).

Smith-Petersen and co-workers² designed the operation of spinal osteotomy in which the spinal column is hyperextended in the lumbar region, enabling the patient to see straight ahead. Smith-Petersen obtained successful results in six patients, all of whom were operated on via a posterior approach. The lumbar spinal column was osteotomized at one, two, or three levels. These osteotomies were confined to the laminae and articular facets and did not involve the vertebral bodies. Thoracic osteotomy was not advised because the thoracic "cage" is also ankylosed and so prevents correction (and the spinal cord, not the cauda equina, is in this region).

Since that time, only a relatively few surgeons have attempted this major operation and most have modified the original technique. La Chapelle³ reported one case in which he used both an anterior and a posterior approach, enabling him to divide the ossified anterior ligaments under direct vision. Herbert⁴ also believed that posterior osteotomy alone was not always sufficient

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Fig. 1. Upper Left, Thirty-five-year-old man with ankylosing spondylitis, characteristic stance before operation. Upper Right, X-ray taken before operation. Lower Left, Corrected stance 6 months after spinal osteotomy. Lower Right, X-ray taken 23 years after operation; correction has been maintained.

to give good correction in patients with considerable ossification of the anterior longitudinal ligament. He reported nine patients, seven of whom required both an anterior and a posterior approach. The anterior osteotomy was performed 2 to 3 weeks after the posterior osteotomy, and "full correction" was obtained at this time. McMaster⁷ found that, in his series of 15 patients followed up for an average of 4 years, all the correction occurred at one level, although the osteotomy was performed at two levels. Despite this, he considered correction at two levels to be safer. However, both Law⁸ and Goel⁷ advocated osteotomy at a single level, and Goel also did not use bone grafts at the osteotomy site. Goel's 15 patients, 11 of whom had ankylosing spondylitis and 4 osteomalacia, were followed up for an average of 2 years 9 months. The largest series reported is that by Law,⁸ who performed 120 osteotomies; he gave no details of the long-term results.

The technique of spinal osteotomy offers a high potential for serious complications. Although all surgeons have reported good early results in patients who did not have serious complications, the long-term benefits are as yet unreported.

The purpose of this paper is to report our experience in spinal osteotomy at the Mayo Clinic, to discuss the complications, and to report the long-term results.

CLINICAL MATERIAL

From 1945 to 1967, 17 patients with ankylosing spondylitis have undergone spinal osteotomy at the Mayo Clinic by one of us (M.B.C.). There were 11 men and 6 women whose average age was 42.2 years, the youngest being 31 years and the oldest 49 years. The average duration of symptoms was 18 years, and all patients had received conservative therapy unsuccessfully. All 17 patients had received physiotherapy, and 7 had worn a back brace. Six patients had irradiation and 10 had received cortisone, all without success.

Other joints affected by the disease in this group of patients were the hips (10 patients), knees (3 patients), and shoulders (3 patients).

All patients wanted surgery because of their grotesque appearance. Only one patient complained of severe pain, and another patient found it difficult to breathe and had discomfort

after eating owing to the pressure of her sternum and ribs on the abdomen.

TECHNIQUE OF OPERATION

The operative technique is basically that described by Smith-Petersen and associates² and only important points will be discussed with reference to our patients.

Endotracheal anesthesia is essential, and great care is taken during intubation not to injure the cervical spinal column which also may be ankylosed. One patient had such a severe flexion deformity of the cervical spinal column that endotracheal intubation was impossible and tracheostomy had to be performed. This patient, because of her severe deformity, could not be placed prone on the operating table and so underwent operation while lying on her side. The remaining patients were all operated on while in a prone position (Fig. 2), the abdomen being left free for adequate abdominal respiration.

The lumbar spinal column distal to the cord termination (L-2) is the preferred site of osteotomy. The lumbosacral level, where exposure of the laminae and facets is difficult, is not desirable either. Thus L3-4 seems the best, although L4-5 or L2-3 can be utilized. Ossification of the ligaments is not important, relative to the decision as to the proper level, because correction occurs after a break in the anterior longitudinal ligament at the site of the intervertebral disk, or as an "open up" fracture of the vertebral centrum, whichever is the weaker.

At the site chosen for osteotomy, a wedge of bone is removed from the adjacent spinous processes and carried anteriorly as an oblique osteotomy cut at an angle of 45° on each side across the ossified ligamentum flavum to the apex at the dura. The lateral portion of the osteotomy is carried through the fused facets into the intervertebral foramina (Fig. 3 left). The important biomechanical point in cutting the osteotomy is to place the transverse axis, about which correction takes place, anterior to the cauda equina. This means extending the osteotomy well forward laterally—including the fused facets—and to the nerve roots. If the osteotomy is cut correctly, the cauda equina will be relaxed as the spinal column is hyperextended; but if the axis of rotation lies posterior to the cauda equina, serious neurologic compli-

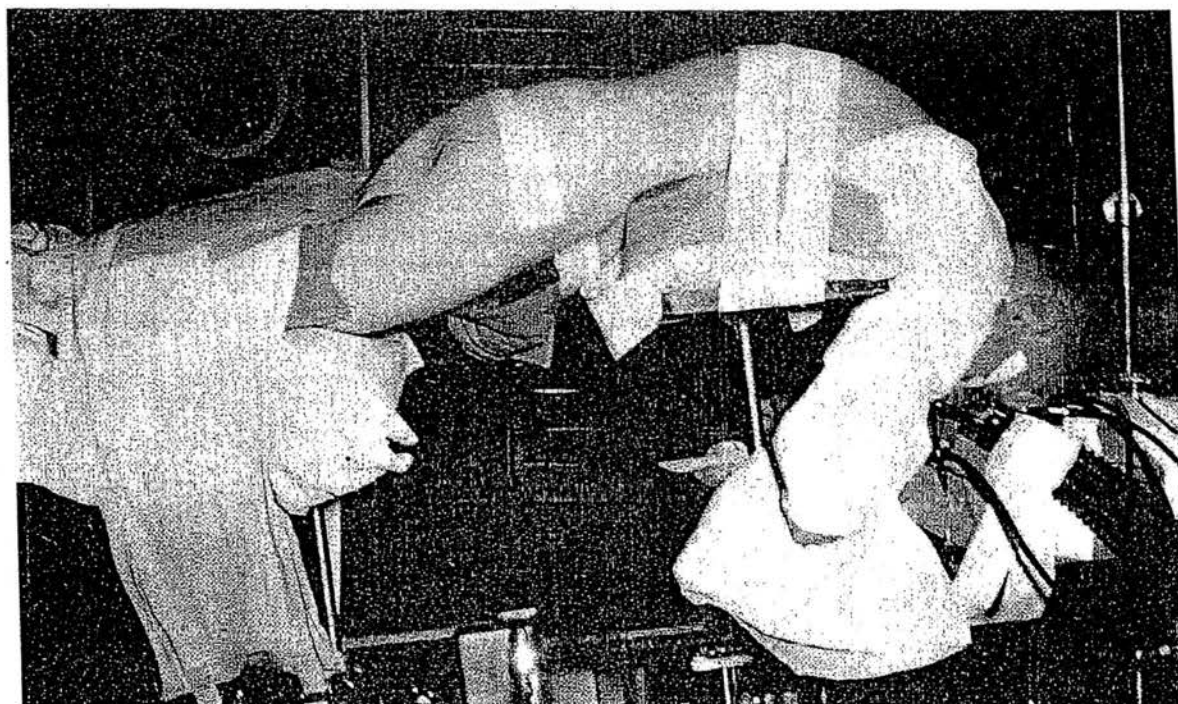


Fig. 2. Position on operating table.

cations may result from stretching of the nerves. The dura is often adherent to the undersurface of the lamina and ligamentum flavum and may be torn. If so, it can be closed by suture, as it was in six of our patients.

The first 11 patients operated on had osteotomies performed at two adjacent levels. But we found, as McMaster⁵ had described, that the correction occurred at one level in all but one patient. With this knowledge the remaining patients were osteotomized only at one level. The levels of the osteotomies are shown in Table 1.

The deformity was corrected as far as possible under direct vision (Fig. 3 *middle*) by raising the head and foot of the operating table. Three patients had a distinct snap as the ossified anterior longitudinal ligament ruptured. In five patients, there was difficulty in closing the osteotomy completely as it tended to spring open. Undue force was not used on these patients, and further correction was obtained in the postoperative period.

Bone flaps were raised from the adjacent laminae, and the chips of bone obtained from the excised wedge of bone were laid across

the osteotomy site (Fig. 3 *right*). Anterior and posterior plaster shells were applied on the operating table while the patient was still anesthetized.

POSTOPERATIVE CARE

All patients were nursed in their anterior and posterior plaster shells for 10 days to 2 weeks, until the wound was well healed. After this period a plaster cast incorporating one thigh was applied, and final correction was obtained.

The technique used on the first 11 patients was similar. Eight of the patients were given a general anesthetic, and three were sedated. The patient was then turned prone on the plaster table, and overhead slings were passed under each shoulder. The patient was hyperextended by raising the shoulders off the table. Once full correction was obtained, the plaster cast was applied. This method gave good correction, but neurologic complications occurred after the procedure in three patients, and the technique was changed.

The last five patients operated on were not anesthetized during cast application. They were given an analgesic and were placed on the Risser frame where the cast was applied with

as much correction as possible. When the cast was dry, hinges were placed laterally, plaster wedges removed posteriorly, and turnbuckles applied anteriorly. The remaining correction was obtained over 1 to 3 weeks by means of the turnbuckles. When full correction was obtained, the cast was repaired. During this period of gradual correction, two patients experienced paresthesias in the legs, but this was only

temporary and soon disappeared after the rate of correction was decreased. This is a much safer way of obtaining the desired correction than by forceful manipulation under general anesthetic either at operation or during cast application, and this is the method we now use and advocate. Neurologic complications, if they occur, can be recognized before they can become serious.

Long-Term Results of Seventeen Patients Undergoing Spinal Osteotomy for Ankylosing Spondylitis

Case	Age (yr); sex	Duration of disease, yr	Level of osteotomy	Degrees of correction	Complications	Follow-up				Outcome
						Duration, yr	Pain	Increasing deformity	Hip deformity	
1	37F	14	L2-3 L3-4	28	Ileus	17	...	Thoracic and cervical spine	...	Died of cor pulmonale after 18 yr
2	43M	18	L1-2 L2-3	35	Sacral sore	24	...	Cervical spine	...	Femoral prosthesis
3	49M	20	L3-4 L4-5	33	...	2	Died of coronary thrombosis after 4 yr
4	35M	16	L2-3 L3-4	48	Weakness in rt arm	23	...	Thoracic and cervical spine	Flexion contracture	Bilateral Judet hip arthroplasties
5	44M	13	L3-4 L4-5	...	Died day after surgery
6	38M	11	L3-4 L4-5	33	...	3.5	Died of coronary thrombosis after 16 yr
7	37F	15	L2-3 L3-4	43	...	13.5	Flexion contracture	Rt total hip arthroplasty
8	49M	20	L3-4 L4-5	36	...	2.5	Flexion contracture	Died of coronary thrombosis after 8 yr
9	31M	12	L3-4 L4-5	20	...	10	Thoracic spine	Thoracic spine
10	40M	25	L2-3 L3-4	40	Ileus	4.25	...	Cervical spine	...	Cervical osteotomy at 1½ yr postop
11	37F	20	L1-2 L2-3	90 reduced to 45	Paraparesis	13	Flexion contracture	Fully recovered from paralysis
12	37F	16	L2-3	30 reduced to 22	Paraparesis	12	No change in paraparesis
13	45F	26	L4-5	37	Temp. paresthesia in legs	11	Thoracic spine	Thoracic spine	Flexion contracture	...
14	47M	25	L4-5	21	Chest infection	3.5	Thoracic spine	Thoracic spine
15	44M	22	L3-4	50	Chronic osteomyelitis; delayed wound healing; chest infect.; ileus	Died 3 mo postop, from acute hemorrhage
16	49M	17	L2-3	48	Delayed wound healing	6.5	Flexion contracture	Rt total hip arthroplasty
17	45F	24	L4-5	45	Ileus; temp. paresthesia in legs; aphasia	5	...	Cervical spine

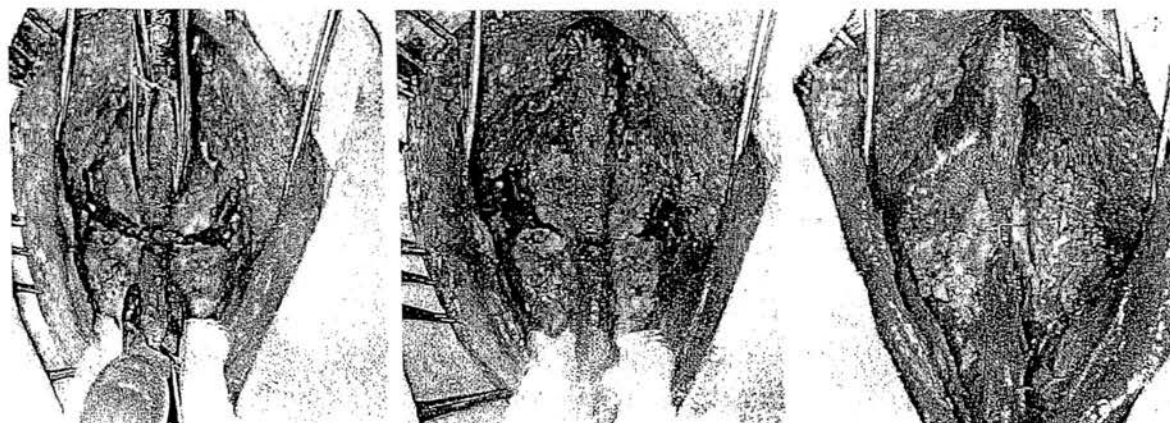


Fig. 3. Operative technique. *Left*, Osteotomy is cut at 45° angle on each side, across ossified ligamentum flavum and fused facets into intervertebral foramina and apices of dura. *Middle*, Osteotomy is closed without force. Cut bone edges are brought together. *Right*, Bone chips are laid across osteotomy site (wire seen here is not usually used).

COMPLICATIONS

Various complications have been reported in the literature after this operation. Law⁶ reported 10 deaths in his series of 120 patients, and McMaster³ had 1 death in his series of 15 patients. Other complications include paraplegia, nerve root irritation, acute dilatation of the stomach, paralytic ileus, superior mesenteric thrombosis, and psychologic disturbances.

In our series, 12 of the 17 patients had complications.

Deaths.—Two patients died in the postoperative period. Both patients were 44-year-old men. One patient died suddenly the day after surgery. Postmortem examination revealed pulmonary edema only and the cause of death remained unknown, although the patient had been receiving cortisone therapy, and the pathologic diagnosis after autopsy was adrenal insufficiency. The second patient died suddenly 3 months after surgery. His early postoperative course was complicated by a chest infection and paralytic ileus, and he continued to have a fever. Postmortem examination revealed chronic osteomyelitis at the site of osteotomy which had slowly eroded a lumbar artery near the aorta, and death was due to hemorrhage of this vessel.

Neurologic Complications.—Five patients had neurologic complications. Two patients, both 35-year-old women, had signs of injury to the cauda equina after cast application. In both, the casts were applied by our original method in which the patients were anesthetized, laid

prone, and hyperextended by means of slings under the shoulders. On awakening from the anesthetic, one patient (case 11) immediately complained of weakness and pain in both legs, although sensation was unimpaired. The cast was removed immediately, but there was no improvement. On dismissal from the hospital after 4 weeks, the patient was walking with crutches. Sensation was normal, but she had generalized weakness of both legs, although the weakness was improving slowly. When seen 2 years later, the patient had no neurologic deficit.

The second patient (case 12) had had a previous lower sacral resection for reasons we could not determine, and, after this, bladder incontinence resulted. On awakening from the anesthetic, she complained of weakness and loss of sensation in the legs; later, she found it difficult to urinate. The cast was removed and the degree of correction reduced, but there was no improvement. On dismissal from the hospital, after several months, the patient still had considerable weakness in both legs. Sensation was unimpaired, except for some hypoesthesia in the perineal area. The bladder problem had been relieved by resection of the bladder neck. Her main complaint was a burning pain in both legs, mainly below the knees. When she was seen 2 years later, her condition was unchanged, but she could walk using long leg braces and crutches. Her condition has remained unchanged.

A third patient (case 4) complained of weakness in his right arm after cast application. This

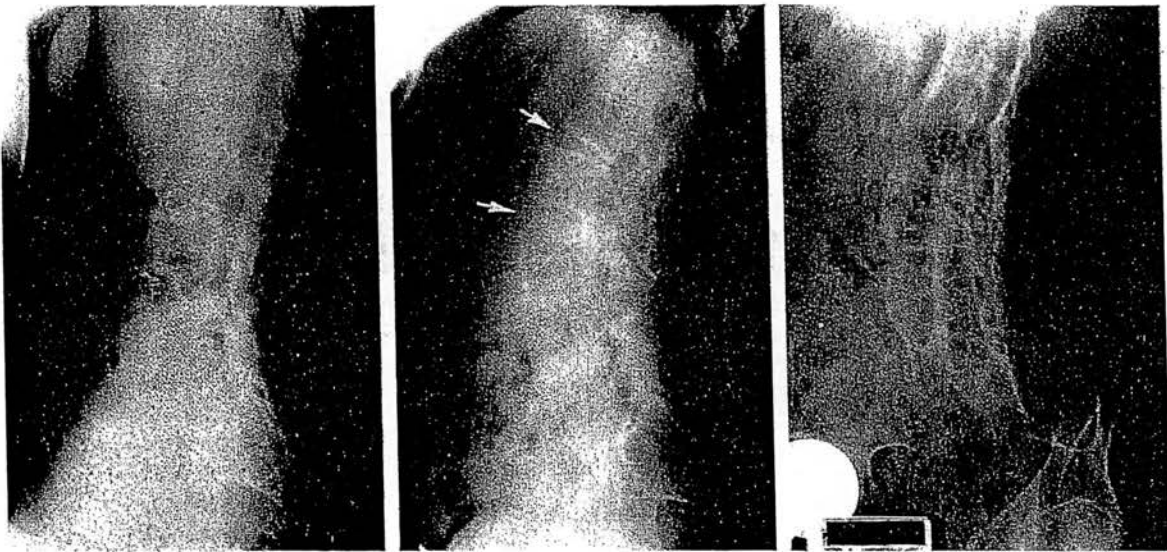


Fig. 4. *Left*, Before operation. *Middle*, One month after operation, showing correction at L1-2 and L2-3 levels due to fracture of ossified longitudinal ligament. *Right*, Twenty-three years after operation.

was caused by pressure of the sling on his axilla. The symptoms disappeared within a few days.

Because of these complications we changed our method of cast application. Two of the five patients on whom this new method was used also had mild neurologic symptoms. These consisted of paresthesia in both legs; the numbness rapidly disappeared when the rate of correction was reduced. We now consider this method of cast application and correction the most suitable and the least dangerous.

Ileus.—Four patients developed paralytic ileus within 48 hours of the operation. In all four, the ileus resolved within a few days.

Aphasia.—One patient became aphasic for 1 hour on the 18th day after the operation. The cause for this was not found, and she has since remained asymptomatic.

Delayed Wound Healing.—This occurred in two patients, one of whom had an associated hematoma. Wounds in both patients healed within a few weeks.

Pulmonary Infection.—Two patients developed pneumonitis—a serious complication in patients whose vital capacity is already reduced. In both, the pneumonitis resolved with treatment.

Sacral Sore.—One patient developed a sacral sore at an area of pressure under his cast; this healed after the cast was trimmed.

POSTOPERATIVE CORRECTION

The usual postoperative program was to maintain the anterior and posterior body cast shells for 10 to 14 days after osteotomy (allowing for turning and body care during this time). Then, a body cast with one extension down the thigh to the knee (single spica) was applied with the patient supine on a Risser frame. Wedging was then begun. When correction was complete, the cast was filled with plaster, and the patient was allowed to ambulate. At 3 to 4 months, the cast was removed and a well-molded body cast without the spica attached was applied and maintained until solid fusion of the grafted area was achieved. The length of time in which a cast was necessary varied from 6 to 15 months, with an average of 7½ months. All patients had bony fusion. When the cast was removed, six of the patients needed to wear a back brace for a few months to 1 year. One patient who complained of back pain because of continued activity of his disease was advised to wear his back brace indefinitely.

Measurements on the roentgenograms, taken after final correction was obtained, revealed corrections that varied from 21° to 50°, with an average of 39°. A commensurate improvement in height and posture was noted. In all but one of the patients with correction at two osteotomy levels, the correction took place at

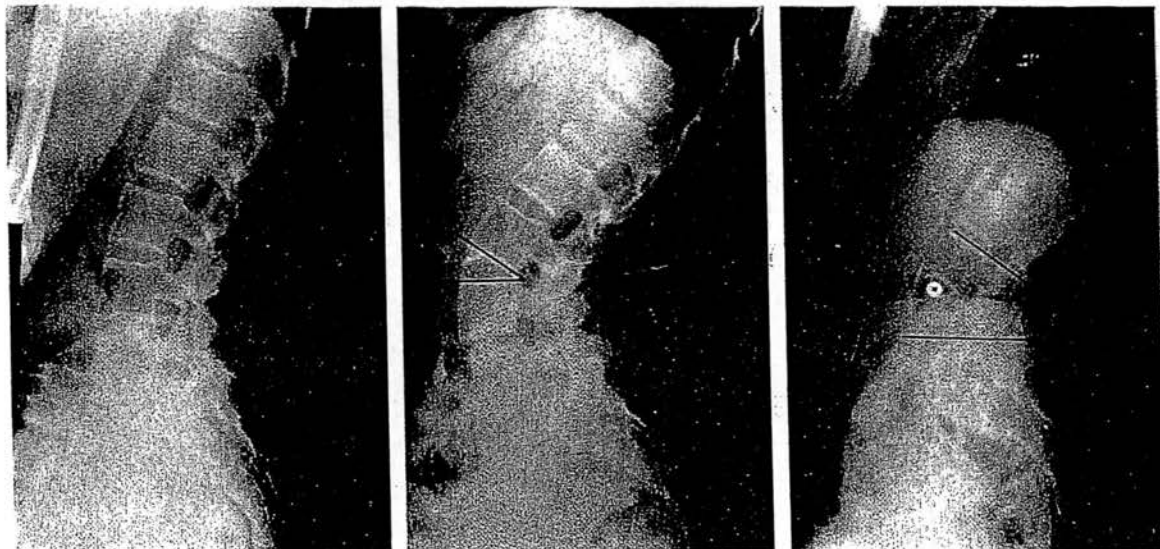


Fig. 5. *Left*, Before operation. *Middle*, Nine months after operation, showing correction at L3-4 level, with beak of bone pulled off lower vertebra. *Right*, Thirteen years after operation. Correction has been maintained.

the lower level. In 13 patients, correction of the deformity resulted in fracture of the anterior longitudinal ligament and separation of the vertebrae anteriorly at the disk level (Fig. 4). In two other patients, a beak of bone was pulled off the lower vertebrae centrum at its anterior superior border (Fig. 5). In the remaining two patients, correction was obtained by splitting of the vertebra transversely and creating an "open-up" fracture (Fig. 6).

The roentgenograms of the two patients with injury to the cauda equina showed that the axis about which correction took place lay posterior to the spinal cord, causing excessive stretch on the cord and cauda equina. In one patient (case 11), 90° of correction was obtained and the vertebrae at the osteotomy site were distracted (Fig. 7 *left* and *middle*). This degree of correction was reduced immediately (Fig. 7 *right*), and the spinal column fused, although there was much loss of correction. In the second patient (case 12), the initial correction was also reduced immediately (Fig. 8 *left* and *middle*) and subsequently fusion took place, with a further slight loss of correction (Fig. 8 *right*).

FOLLOW-UP

Excluding the two patients who died in the post-operative period, the remaining 15 patients have been followed up for periods that varied from

2 to 24 years, with an average of 10 years. During this follow-up period, four patients died: three from coronary thrombosis at 4, 8, and 16 years after surgery, and 1 from cor pulmonale 18 years after surgery. The final follow-up for these patients is taken as the last time they were seen and examined.

Of the two patients with neurologic motor complications, one (case 11) made a complete recovery within 1 year. She has been followed up for 13 years and feels that her operation is a success as she is able to do her housework and care for her three children. The second patient (case 12) has been followed up for 12 years. She still has some generalized weakness in her legs which require long leg braces. There is no obvious sensory loss although she complains of a burning sensation in both legs below the knees. Her back is not painful, providing she wears a back brace.

All of the patients were pleased with their operations, which now enabled them to see ahead and which improved their appearance and increased their height (Fig. 1 *right*). The emotional benefit is great. Two patients had further benefits from the operation. A 49-year-old man who was unable to have sexual relations prior to surgery later noticed a marked change in his married life and was able to father a child. Another patient who had a severe hematemesis

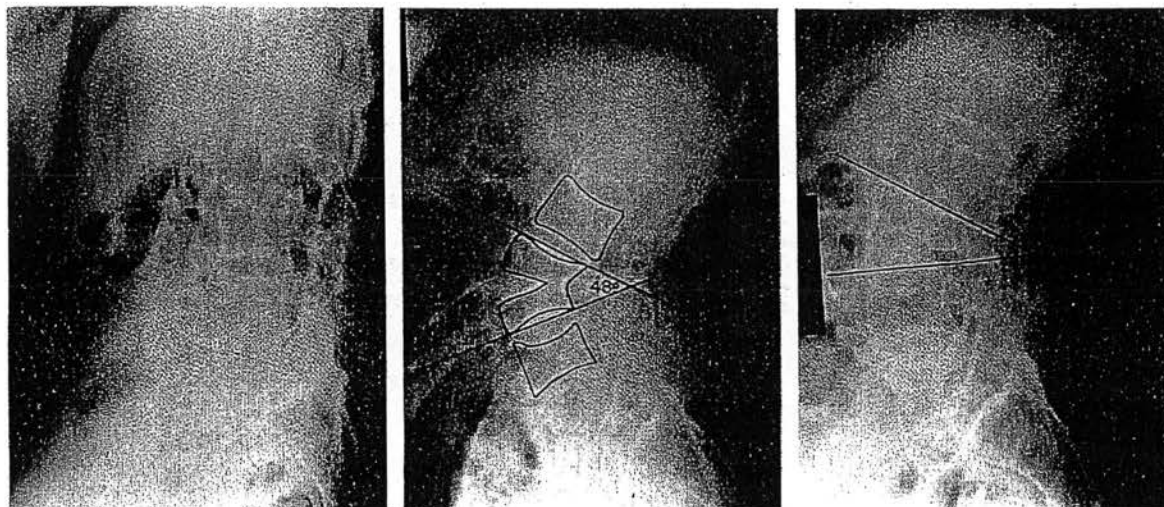


Fig. 6. *Left*, Ten weeks before operation. *Middle*, Five months after operation, showing the "open-up" fracture in L-4. *Right*, Six years later. "Open-up" fracture has now filled in with bone. Loss of 10° in correction at osteotomy site.

underwent emergency abdominal surgery in which there was no difficulty in the incision being made or the abdominal contents being exposed.

Three patients continued to have back pain; the pain was in the upper thorax and cervical spinal regions where the disease was still active. No pain was felt at the site of the osteotomy.

Careful measurement of the roentgenograms

of all the patients after their final cast removal and during follow-up revealed that only three had decreases in the degree of correction at the osteotomy site. These decreases varied from 6° to 12° and occurred following removal of the cast, after which no change was noted. Decrease was probably caused by removal of the cast before complete bony fusion occurred at the osteotomy site. Eight other patients noticed

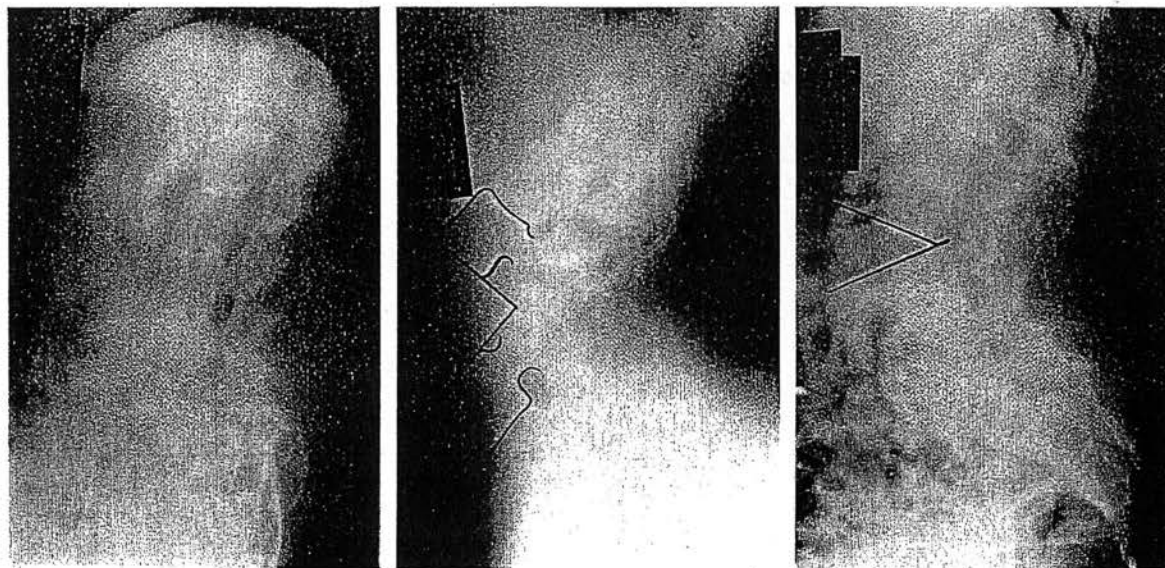


Fig. 7. *Left*, Before operation. *Middle*, Immediately after correction under general anesthetic. Correction of 90° at L3-4 level. Vertebrae have been distracted, and axis of correction lies posterior to cauda equina. *Right*, Six weeks after initial degree of correction was reduced.

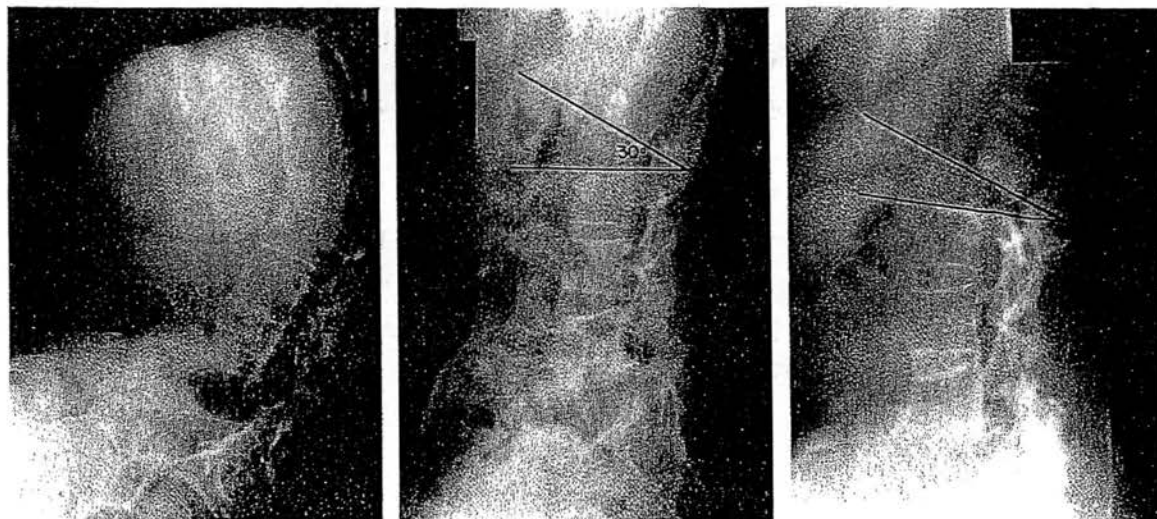


Fig. 8. *Left*, Before operation. *Middle*, Six months after operation, showing that osteotomy is fused but axis of correction lies posterior to cauda equina. *Right*, Twelve years after operation.

deterioration in their spinal deformity after cast removal. This increasing deformity occurred slowly over the years and affected the thorax and cervical spinal regions and was due to a continuation of the disease process in the areas not yet ankylosed by bone. One of these patients later required a cervical osteotomy, and another patient noticed 6 years after her surgery that she had lost 8 inches in height because of a combination of increasing thoracic kyphosis and flexion contracture of the hips, although the degree of correction in the lumbar spinal area had remained stationary. Only one of the eight patients thought that his surgery had not been beneficial, and this was because of continuing pain in the thoracic spinal region.

During the follow-up period, seven patients developed increasing symptoms in joints other than in the spinal column. Six complained of pain and stiffness in their hips and increasing flexion contracture, which bent them over farther and detracted from the results of spinal surgery. One patient had bilateral Judet arthroplasties 3 years after spinal surgery and still has hip complaints at 23-year follow-up. Another patient had a femoral prosthesis inserted at 5 years, and at 12 years this was replaced by a Charnley total hip. He now has no hip complaints (at 13½-year follow-up, 18 months after total hip arthroplasty). The third patient had a cup arthroplasty after 1-year follow-up which re-

quired revision 1 year later and finally a Charnley total hip replacement. He now has no complaints (at 13-year follow-up, 15 months after total hip arthroplasty). A fourth patient has had a femoral prosthesis inserted. The remaining three patients with hip problems all received physiotherapy and conservative treatment.

Other joints that became symptomatic were the shoulders (three patients) and knees (one patient). All were treated conservatively by heat, physiotherapy, and cortisone injections.

DISCUSSION

All previous studies (as well as our present one) have shown that, although spinal osteotomy is a potentially dangerous operation with occasional serious complications, most patients obtain early good results and benefit both physically and psychologically.

Neurologic complications can be prevented by careful operative technique and good post-operative care. The osteotomy is cut carefully, with the axis of correction anterior to the cord. As much correction as possible is obtained on the operating table, but undue force should not be used if the osteotomy does not close properly. We find that safe further correction can be obtained afterward by means of a turnbuckle cast, and any sign of cauda equina stretching or pressure can be detected early.

In this study, we have attempted to determine

if the good early results of spinal osteotomy are permanent. We know that ankylosing spondylitis is a continuing process, with exacerbations and remissions until eventually the disease "burns itself out," at which time the spinal column may or may not be completely ankylosed. Usually, the lumbar spinal region is affected first, and the thoracic and cervical regions are affected later. Spinal osteotomy is just one incident in this continuing process. We have found that the good initial correction in the lumbar region *remains* once the osteotomy is fused, but that if the disease is still active in the thoracic and cervical regions, or in the hips, these continue to deform and detract from the initial correction. If spinal osteotomy is performed near the end of the natural course of the disease or after it is "burned out," a more lasting correction probably is possible, because there is less tendency for the deformity to progress above the osteotomy site. Unfortunately, it is not always possible to wait until the disease is quiescent if there is already a crippling spinal deformity. In these patients, some deterioration should be expected after osteotomy; and a brace, exercises, and other measures should be

used after the cast is removed.

Careful preoperative examination should determine whether the flexion deformity of the patient is due to the spinal column, the hips, or both. Forty-two percent of patients have hip involvement,¹ and an increasing hip flexion contracture also will detract from the improved stance provided by spinal osteotomy. Since the advent of total hip replacement, many of the hip problems in this disease can be solved. Should a hip flexion contracture develop after spinal osteotomy, hip surgery will often restore the patient to an upright stance.

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A TECHNIQUE FOR LUMBAR SPINAL OSTEOTOMY IN ANKYLOSING SPONDYLITIS

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Fourteen patients with ankylosing spondylitis had an extension osteotomy for severe flexion deformity of the spine. The Smith-Petersen technique was modified by using a compression device which allows a slow, finely controlled closure of the osteotomy, and provides rigid internal fixation. There were no serious neurological complications. All the patients were able to see straight ahead after operation, and all had solid fusion at nine months, having maintained good correction.

Ankylosing spondylitis may sometimes cause one of the most disabling deformities seen by the orthopaedic surgeon. The characteristic spinal deformities are flattening of the normal lumbar lordosis, and an increasing smooth thoracic kyphosis with the head and neck thrust forwards. Occasionally there is increasing flexion at the cervicothoracic junction. Eventually the whole spine undergoes bony ankylosis in this deformed position, and the patient is bent forward and forced to look down at the ground. This ugly posture is not only functionally disabling but is also psychologically disturbing.

Smith-Petersen, Larsen and Aufranc (1945) recognised the plight of these unfortunate patients and devised the operation of spinal osteotomy, by which the spine is hyperextended in the lumbar region, enabling the patient to see straight ahead. Since their paper relatively few surgeons have attempted the operation (La Chapelle 1946; Adams 1952; Herbert 1959; Law 1959; McMaster 1962; Goel 1968; McMaster and Coventry 1973). Nearly all of these have reported a high incidence of major complications. The reported mortality rate has varied from 8% to 10% and neurological complications, up to and including paraplegia, have occurred in as many as 30% of the patients. Other complications have included rupture of the aorta, acute dilatation of the stomach, superior mesenteric thrombosis and psychological disturbances. Only Simmons (1977), in a study of 19 patients, reported no deaths or neurological complications following lumbar spinal osteotomy. He attributed this success to performing the operation under local anaesthesia, thus allowing continuous monitoring of the neurological state.

This paper reports the results of a prospective study of methods designed to reduce the incidence of neurological complications during and after lumbar spinal osteotomy. A modification of the usual technique was used and angular correction was obtained with a specially-designed device which allowed slow and finely controlled closure of the osteotomy, as well as providing rigid internal fixation.

CLINICAL MATERIAL

From June 1980 to August 1982, at the Princess Margaret Rose Orthopaedic Hospital, Edinburgh, 14 patients with ankylosing spondylitis each had an extension osteotomy of the lumbar spine to correct a severe flexion deformity. All the operations were performed by the author using a special method of intra-operative correction and internal fixation. There were 11 men and three women whose mean age at operation was 42 years (range 31 to 66 years). Their mean duration of symptoms was 21 years (range 6 to 44 years) and all had been treated conservatively without success.

Two patients had become much more stooped because of vertebral compression fractures of L1 and L2 respectively, which they had sustained in falls. Neither had any neurological abnormalities and the fractures healed satisfactorily. Two other patients developed increasing pain in the back and noticed that their flexion deformity was getting worse. Both had developed a spontaneous pseudarthrosis in their ankylosed spine, one between T12 and L1, and the other between L1 and L2.

In seven of the patients the hips also were affected and five of these had had bilateral total hip replacement 3 months to 10 years before spinal osteotomy. These hip replacements were needed to correct fixed flexion and to relieve pain.

Assessment of the deformity. The main indication for the operative correction of a severely flexed posture in a patient with ankylosing spondylitis is the patient's inability to see ahead for more than a few feet. It is,

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however, important to assess the contribution of all levels of the spine and of the hips to the flexed posture before attempting correction. Severe flexion contracture of the hips can often be corrected by soft-tissue release and total hip replacement (Bisla, Ranawat and Inglis 1976; Williams *et al.* 1977) and this may be sufficient by itself to allow the patient to see straight ahead.

A spinal osteotomy is indicated only if the hips are not significantly deformed or if, after hip operations, the patient is still unable to see ahead (Case 11, see Figs 12, 14 and 16). The thoracic spine is usually the most flexed region but osteotomy at that level does not help because the ankylosed thoracic cage prevents extension of the spine. A thoracic deformity is best overcome by the creation of a compensatory lumbar lordosis by means of a lumbar osteotomy (Figs 1 and 2). The safest level to perform the osteotomy is in the lumbar spine distal to the first lumbar vertebra because at this level there is a relatively spacious vertebral canal containing the cauda equina, which is less easily injured than the spinal cord. Ideally the lumbar spine should be extended until the patient can see straight ahead. The centre of gravity of the upper body should then lie just behind the site of the osteotomy, helping to maintain correction.

In a few patients the major flexion deformity is at the cervicothoracic junction. Correction of this deformity by a lumbar osteotomy could possibly unbalance the patient and still not enable him to see straight ahead (Figs 3 and 4). These patients require an extension osteotomy of the cervical spine (Simmons 1977).

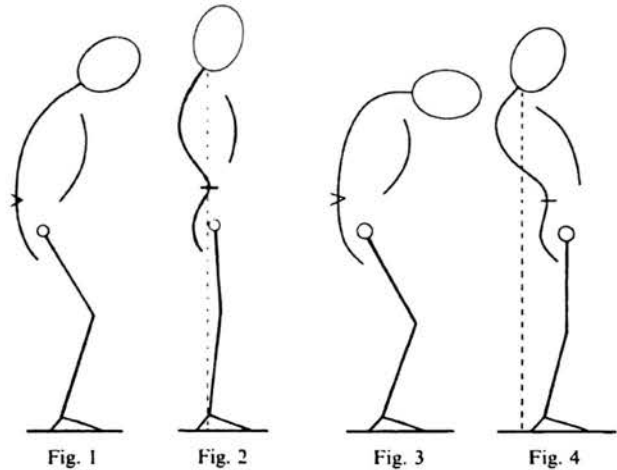
TECHNIQUE OF OPERATION

Endotracheal anaesthesia is essential. Great care is necessary during intubation to avoid accidental injury to the possibly ankylosed cervical spine. Neck stiffness and deformity make intubation very difficult and the use of a fibre-optic laryngoscope is often necessary if tracheostomy is to be avoided.

The patient is turned on to the operating table with care to avoid injury to the ankylosed neck, and positioned face down, with the chest and pelvis supported on special firm foam blocks which leave the abdomen hanging free. This position reduces intra-abdominal pressure; combined with hypotensive anaesthesia and infiltration of the operation site with a 1:400 000 solution of adrenaline, it greatly reduces bleeding during operation and makes it easier to identify the neural structures (Malcolm-Smith and McMaster 1983).

The lumbar spine is exposed through a midline incision extending from T12 to L5. The paraspinal muscles are stripped subperiosteally from the bone laterally to the tips of the transverse processes at the level of the osteotomy. The anatomical details are usually obscured by ossification of the interspinous ligaments, the ligamentum flavum and the interfacetal joints.

The technique of the osteotomy is basically that described by Smith-Petersen *et al.* (1945), and modified



Figures 1 and 2—Flexion deformity in the thoracolumbar spine can be overcome by creating a compensatory lumbar lordosis. Figures 3 and 4—If the major flexion deformity is at the cervicothoracic junction, an attempt at correction by a lumbar extension osteotomy may unbalance the patient.

by Adams (1952) and Law (1959) (Figs 5 and 6). It was not found necessary to perform an anterior spinal osteotomy as a separate stage as described by Herbert (1959) and La Chapelle (1946). The preferred site for the posterior spinal osteotomy is either between the second and third lumbar vertebrae or between the third and fourth. These sites are distal to the spinal cord and are far enough from the sacrum to allow application of the internal fixation device. Identification of the level is made by noting the position of the last rib or by counting the vertebrae up from the sacrum.

The bony sites for the application of the internal fixation device must be prepared before cutting the osteotomy. With an osteotome, notches are carefully cut in the ossified ligamentum flavum on either side of the spinous processes of the vertebrae above and below the site of the osteotomy. Initially these notches are made only in the outer cortex of the bone and are completed by hammering a sharp Harrington hook (No. 1251) on an introducer until the hook lies firmly within the bone. The upper two hooks should lie over the top of the lamina of the vertebra at the level above the osteotomy site, and the lower two hooks around the bottom of the lamina at the level below the osteotomy site. Once each bony site is prepared for later use the sharp hook is removed. Hammering the hooks after the osteotomy has been cut could displace the vertebrae and cause neural damage.

The osteotomy removes a posteriorly based wedge of bone which includes the adjacent spinous processes and the ossified interspinous ligament; it exposes the interlaminar space. The ossified ligamentum flavum in the midline is nibbled away with rongeurs until a small area of dura is exposed. The dura is frequently adherent to the undersurface of the ossified structures; it can easily be torn unless it is separated carefully with a fine blunt dissector. Once separated, the dura is widely

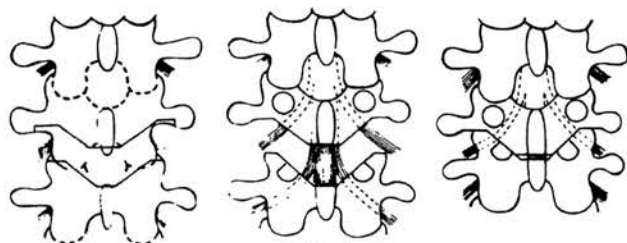


Fig. 5

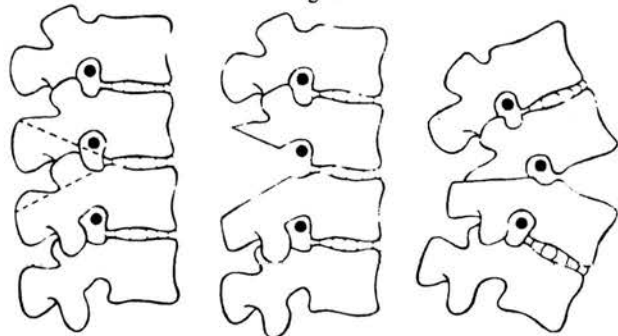


Fig. 6

Diagrams of three stages of the operation to show the recommended technique for extension osteotomy of the lumbar spine.

exposed in the interlaminar space, using laminectomy rongeurs to remove bone. The fused interfacetal joints at the same level are identified and the removal of bone is carefully extended from the midline across these joints at an angle of 45° on each side (Figs 5 and 6). Bone is removed on each side to leave a slot extending anteriorly through the fused facet joint into the intervertebral foramen. The thickness of the bone removed is often between 1 and 2 cm. The bony sides of the osteotomy defect should be parallel and up to 8 mm apart, depending on the angle of correction required. During the preparation neural structures are protected with a blunt dissector and overlying bone is removed with fine osteotomes and rongeurs. It is very important to ensure that the transverse axis of eventual closure, about which correction will occur, lies anterior to the cauda equina. This means that the osteotomy must extend anteriorly so that its apex lies at the anterior margin of the intervertebral canal, and laterally to give good exposure of the nerve roots (Figs 5 and 6).

If the osteotomy is cut correctly, the cauda equina will be relaxed as the spinal column is extended and the osteotomy closes; an incorrect cut will allow hinging on the bony structures behind the neural elements, and result in serious neurological complications from stretching of the cauda equina. It is also important to remove enough bone from the pedicles above and below the intervertebral foramina to ensure that the nerve roots are not pinched as the osteotomy is closed.

Once the osteotomy has been completed, no attempt is made to close the wedge until the internal fixation device has been applied. It was found to be very difficult to insert the Harrington hooks into the prepared fixation sites when they were already mounted on a

compression rod. Blunt Harrington hooks (No. 1253) were therefore modified in the Bio-Engineering Laboratories at the Princess Margaret Rose Hospital. A slot was cut in the top of the hook so that it was able to accept a compression rod after the hook had been placed in the bone. Similar hooks are now available commercially but these are too slim and tend to cut through the relatively soft bone found in these patients. The Harrington No. 1253 hook has a broader shoe and is therefore less likely to cut through bone when compression is applied.

Four of these modified hooks are inserted into the prepared fixation sites above and below the osteotomy. A Harrington compression rod is pressed down into the hooks on each side of the spine. These rods are held in place by special bushes which are advanced along the rods and into the base of each hook where they are enclosed by the hook but cannot pass through it. The osteotomy is closed by slowly tightening the nuts on the threaded rods, thus applying compression by the four hooks.

The advantage of this technique is that it allows a slow and finely controlled closure of the osteotomy; there is no sudden snap or rapid closing of the wedge space. As the osteotomy closes, the dura, which is well exposed in the midline, is seen to wrinkle, confirming that the neural elements are being relaxed. If the dura does not wrinkle, the osteotomy should be inspected to make sure there is no premature impingement of bone posterior to the neural structures. The interlocking 'V' shape of the osteotomy makes it very stable and prevents any rotation. The compression rods prevent distraction and maintain correction.

Once the osteotomy is closed a wake-up test is performed and the neurology of the lower limbs is checked. If there is a neurological deficit the fixation can be released. If all is well, bone chips removed during the osteotomy are placed posteriorly and also on both sides between the transverse processes of the vertebrae above and below the osteotomy. These transverse processes are nearer to each other once the osteotomy has been closed (Fig. 5).

Postoperative care. Gastric dilatation is a serious complication which may occur in the few days after lumbar spinal osteotomy. Extension of the spine may cause the third part of the duodenum to be pinched between the superior mesenteric vessels which pass over it and the spine which lies behind it. The obstruction usually resolves after a few days but if it is not recognised the stomach may become distended and the patient may vomit large quantities of fluid, with the danger of aspiration into the lungs. Because of the ankylosed thoracic cage, these patients cannot cough properly and easily develop aspiration pneumonia, which can be fatal. To prevent this complication, a nasogastric tube is passed before the patient wakes up from the anaesthetic and is in place for 48 hours or until it is apparent that there is no obstruction.

After the operation, the patient is nursed on a firm bed with a small pillow under the osteotomy site. Because of the rigid internal fixation, external support is unnecessary, and the patient can be log-rolled safely from side to back to side. Physiotherapy is needed to encourage coughing and the removal of pulmonary secretions. The use of an absorbable subcuticular stitch allows the application of a well-moulded underarm plaster jacket five days after the operation. This jacket is applied with the patient lying face down on a plaster frame and no further correction is attempted. The patient is then allowed to walk and is usually fit to leave hospital two weeks after operation. After a few weeks, light sedentary work is allowed but heavy lifting is forbidden until the spine is solidly fused. The plaster jacket is removed after nine months provided that radiographs show a solid fusion. A Jewett brace is worn for a further three months and after this there are no restrictions.

Complications. There were no deaths or neurological complications in any of the 14 patients treated by this technique.

The only complication during operation was a small tear in the dura in three patients. This occurred in each

case while the ossified ligamentum flavum was being nibbled away from the midline before the osteotomy was extended. Dura was adherent to the ligamentum flavum and could not be separated until an initial small opening had been made in the ossified ligament. The small tears were repaired and gave no further problems.

Soon after operation two patients had excessive volumes of bile-stained fluid aspirated through their nasogastric tubes. In both patients, bowel sounds were normal and there was no other evidence of paralytic ileus. Obstruction of the third part of the duodenum was suspected, but gastric distension was not allowed to develop, and the gastric aspirations settled within a few days in both patients, allowing removal of the nasogastric tubes without further complications.

RESULTS

The 14 patients have been followed-up from one year to three years, with a mean of one year eight months (Table I). Osteotomy was between the second and third lumbar vertebrae in nine patients and between the third and fourth lumbar vertebrae in three. The two patients who had a spontaneous pseudarthrosis before operation had their osteotomy performed at the site of the

Table I. Details of 14 patients with ankylosing spondylitis who underwent extension osteotomy of the lumbar spine

Case	Age (years)	Sex	Before spinal surgery			Complications	Correction of curve		Duration of follow-up (years)	Back pain		
			Duration of disease (years)	Condition of hips	Level of osteotomy		Soon after operation (degrees)	Final follow-up (degrees)		Before	After	Increase in deformity
1	43	F	26	Bilateral THR at 36 years	L3-L4	Dural tear	44	44	3	Severe	None	Slight flexion at cervicothoracic junction
2	35	M	10	—	L2-L3	—	40	38	2.5	Mild	None	
3	66	F	44	—	L3-L4	Duodenal obstruction	30	30	2	Severe	Mild	
4	31	M	7	20 flexion deformity	L2-L3	—	33	31	2.5	Severe	Mild	Hip flexion contractures Required bilateral THR at age of 33
5	58	M	41	30 flexion deformity	L2-L3	—	44	34	2.2	None	None	
6	47	M	20	Bilateral THR at 38 years	L2-L3	—	40	33	1.9	Mild	None	Slight flexion at cervicothoracic junction
7	40	M	19	Bilateral THR at 39 years	T12-L1 (pseudarthrosis)	—	30	20	1.5	Severe	None	
8	35	F	16	Bilateral THR at 25 years	L3-L4	—	37	32	1.5	Mild	None	
9	35	M	18	—	L2-L3	—	35	35	1.8	Severe	None	
10	47	M	30	—	L2-L3	—	37	25	1.4	Severe	None	
11	31	M	6	Bilateral THR at 31 years	L2-L3	Dural tear	48	44	1.5	None	None	
12	46	M	23	—	L2-L3	Dural tear	45	45	1.6	Mild	None	
13	37	M	15	—	L1 L2 (pseudarthrosis)	—	26	20	1.6	Severe	None	
14	33	M	15	—	L2-L3	Duodenal obstruction	38	30	1	Moderate	None	

THR, total hip replacement

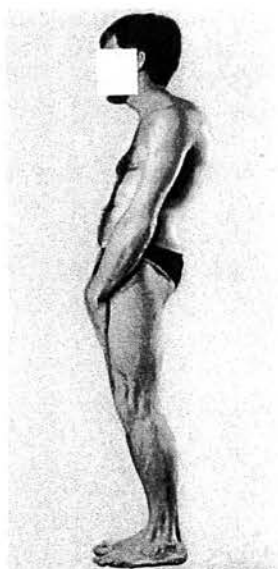
pseudarthrosis. This was between T12 and L1 in one patient and between L1 and L2 in the other.

Radiographs of the spine after operation showed that, as the posterior wedge closed, correction occurred in the anterior part of the vertebral column in one of three different ways. In 10 patients there was a fracture of the ossified anterior longitudinal ligament with opening of the anterior disc space (Case 2, Figs 7 to 11; Case 11, Figs 12 to 17). In two patients the anterior longitudinal ligament avulsed a bone fragment from the lower of the two vertebrae. In the other two patients, the anterior longitudinal ligament remained intact and there was a wedge compression fracture of the posterior part of one of the vertebral bodies at the level of the osteotomy (Case 4, Figs 18 and 19). The immediate postoperative correction measured on the radiographs of the spine ranged from 26° to 48° with a mean of 38°.

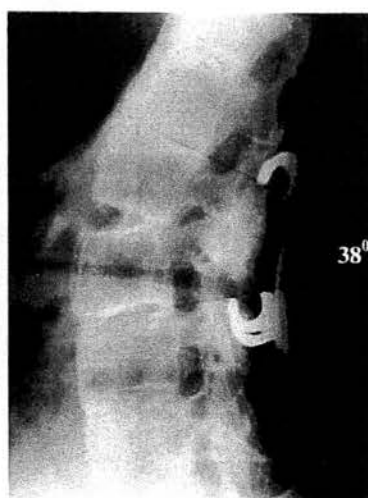
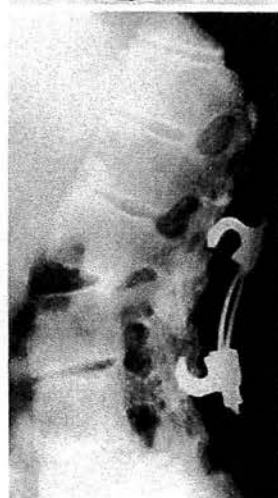
All patients had solid posterior fusion on radiographs taken when the plaster jacket was removed after

nine months. One patient had a broken Harrington rod but the rod on the other side was intact and there was minimal loss of correction. After fusion of the osteotomy, there was very little loss of correction in any patient. At final follow-up, the mean correction at the osteotomy site was 33° (range 20° to 45°) showing a mean loss since operation of 5° (range 0° to 12°).

The osteotomy had corrected all the patients sufficiently for them to see straight ahead (Case 2, Figs 7 and 9; Case 11, Figs 12 and 16). Both their height and their posture was significantly improved and all were pleased with the result. During the period of follow-up, satisfactory correction was maintained at the osteotomy site, but three patients noticed a deterioration in their posture caused by increasing flexion deformity at other sites. Two of these patients had increasing flexion at the cervicothoracic junction, not yet severe enough to require treatment, though they may need cervical osteotomy in the future. The third patient had increasing



Case 2. Figures 7 and 8—A 35-year-old patient with a severely flexed posture due to thoracic kyphosis. Figures 9 and 10—One year after extension osteotomy at L2-3. The spine is solidly fused and the patient can easily see straight ahead. Figure 11—Two-and-a-half years later good correction has been maintained.



Figs 7 and 8

Figs 9 and 10

Fig. 11

pain and flexion contractures at the hips. His posture was restored by bilateral total hip replacements 18 months after his spinal osteotomy. At final follow-up all the patients could still see straight ahead.

Before their osteotomies, all but two of the patients had had back pain. Seven of these 12 patients, including the two with spontaneous pseudarthrosis, described their pain as being very severe, while five had mild to moderate pain. At final follow-up only two patients complained of mild backache.

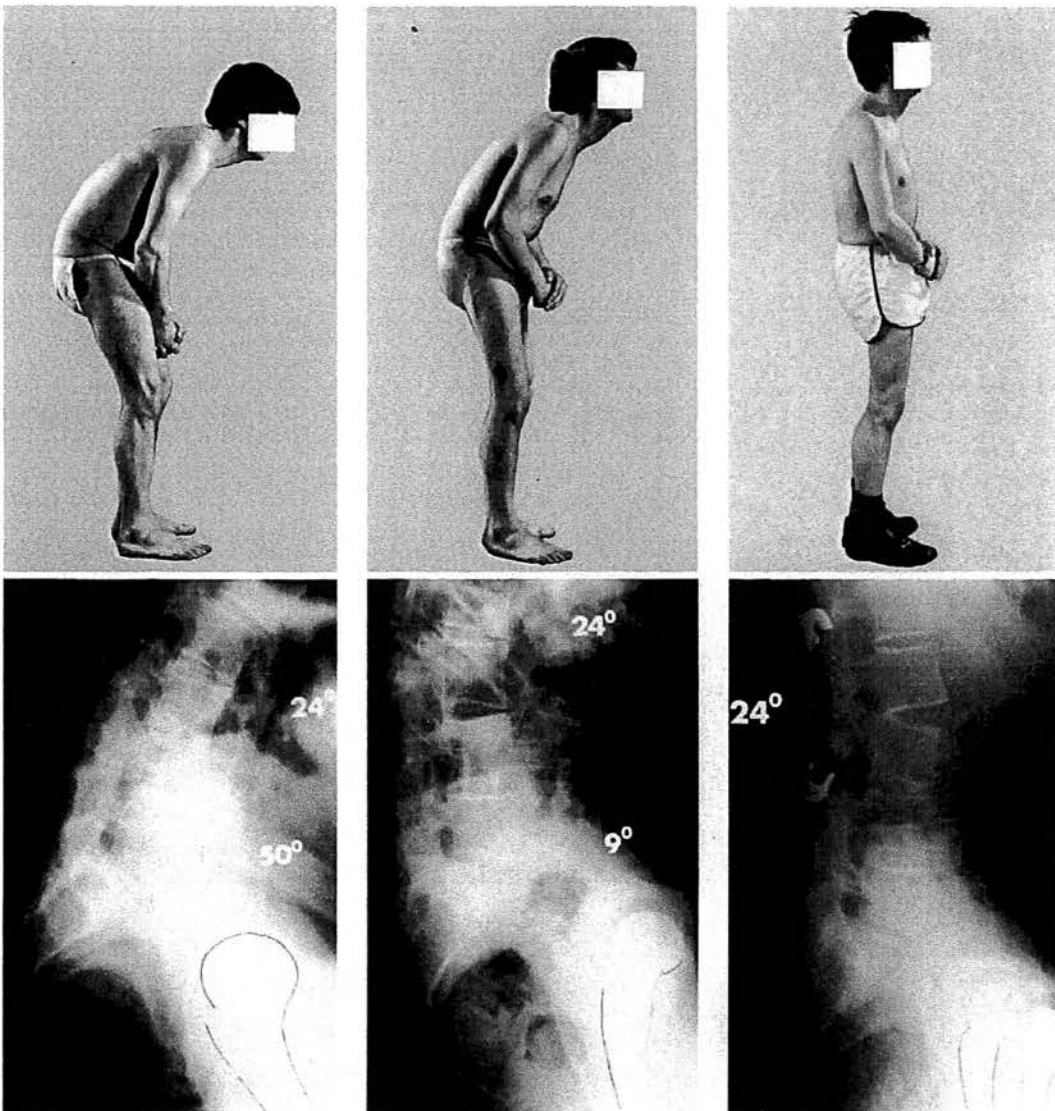
Before spinal osteotomy, deformity had prevented all but two of the 11 men from working. After the operation, seven of these men were able to return to work after recovery periods varying from three months to one year. Of the women, two were housewives and one

had retired; all three felt that their operation enabled them to work more easily in the home.

A further major advantage of the operation, which was expressed by all the patients, was an improvement in self-esteem. Before the osteotomy many of these patients, especially the women, were reluctant to be seen outside their homes. The new appearance provided a marked psychological improvement.

DISCUSSION

This study has shown that modification of the operative technique and the method of correction make lumbar spinal osteotomy for severe deformity in ankylosing spondylitis possible, without the high incidence of major complications which has been reported in other series.



Figs 12 and 13

Figs 14 and 15

Figs 16 and 17

Case 11. Figures 12 and 13—This 31-year-old patient with a severely flexed posture caused by 50° flexion contractures of both hips and a lumbar kyphosis of 24°. Figures 14 and 15—After bilateral hip replacements the flexion contractures were reduced to 9° but the patient still had difficulty in seeing ahead. Figures 16 and 17—Extension osteotomy was performed at L2-3 and the spine corrected by 48° to give a 24° lumbar lordosis. The patient stands upright and can see straight ahead.

All these series used the technique described by Smith-Petersen *et al.* (1945), in which the spine is osteotomised through the posterior elements and corrected by direct pressure on the osteotomy site while the upper body and legs are extended.



Fig. 18



Fig. 19

Case 4. The spine has been corrected 31° by extension osteotomy. There is a posterior wedge compression fracture of L2.

The ossified anterior vertebral column is fractured by this pressure. This often occurs with a sudden snap, making it difficult to prevent displacement and over-correction, which could result in serious neurological complications (McMaster and Coventry 1973). The neural structures can also be stretched by this extension if the osteotomy has been incorrectly cut and the axis of angulation lies either in the same plane as the cauda equina or posterior to it. A third source of problems may be the nipping of nerve roots in their intervertebral canals at the level of the osteotomy when too little bone has been removed. After the closure of the wedge osteotomy using the old technique, the spine becomes very unstable and needs to be controlled by the application of plaster shells, before the patient is removed from the operating table.

By contrast, the technique described in this paper allows a slow and finely controlled closure of the osteotomy without any sudden displacement of the vertebrae. During the correction, the dura can be seen to wrinkle, indicating that neural structures are being relaxed and that the osteotomy has been cut correctly. If this does not occur, compression can be released and more bone removed. Up to 45° of correction can be obtained safely.

The combination of the 'V' shape of the osteotomy and the compression apparatus provides immediate rigid and stable fixation, which allows the patient to be nursed safely in an ordinary bed without external support. It also facilitates the respiratory care of these patients, who

have difficulty in coughing because of their rigid chests.

We had no serious complications using this technique, and all 14 patients were corrected sufficiently to see straight ahead. All had a solid fusion in nine months with maintenance of good correction at the osteotomy, though it should be noted that the overall posture in three of our patients deteriorated during follow-up because of increasing flexion at either the cervicothoracic junction or the hips. McMaster and Coventry (1973), reporting on 17 patients followed for a mean of 10 years after lumbar osteotomy, found that once the osteotomy had fused correction in the lumbar region was maintained; but active disease in the thoracic and cervical spine or the hips could allow increased local deformity and detract from the initial overall correction. The correction was more lasting if osteotomy was done after the disease had "burnt out". Unfortunately, if the spinal deformity is already crippling, it is not possible to wait for the disease to become quiescent. However, correction lost at sites other than that of the lumbar osteotomy can often be improved by either hip replacement or cervical osteotomy.

In conclusion, lumbar spinal osteotomy is a potentially dangerous operation but, with carefully controlled correction at operation and rigid internal fixation, many of the complications can be overcome. The operation provides not only major functional advantages but also psychological benefits.

I am grateful to Dr Mark Coventry who first introduced me to the Smith-Petersen technique of lumbar spinal osteotomy while I was working at the Mayo Clinic, Rochester, Minnesota, USA.

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OSTEOTOMY OF THE CERVICAL SPINE IN ANKYLOSING SPONDYLITIS

MICHAEL J. McMASTER

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Fifteen patients with ankylosing spondylitis who had developed a severe flexion deformity of the cervical spine which restricted their field of vision to their feet, were treated by an extension osteotomy at the C7/T1 level. The operation was performed under general anaesthesia with the patient in the prone position and wearing a halo-jacket. Three had internal fixation using a Luque rectangle and wiring. Their mean age was 48 years.

Before operation the mean cervical kyphosis was 23°; this was corrected to a mean of 31° of lordosis, a mean correction of 54°. All the patients were able to see straight ahead. One patient with normal neurology soon after operation became quadraparetic after one week; two others had unilateral palsy of the C8 root, which improved. There was subluxation at the site of osteotomy in four patients, and two of them developed a pseudarthrosis which required an anterior fusion.

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Ankylosing spondylitis may produce an extreme flexion deformity at the cervicothoracic junction. The patient is unable to see straight ahead and the proximity of the chin to the chest may interfere with eating. This posture is functionally and psychologically disturbing for the patient who may confine himself to the home. The only available treatment is an extension osteotomy of the cervical spine.

In 1958, Urist¹ described a patient with ankylosing spondylitis and a severe cervical kyphosis which was corrected by removing a posterior wedge of bone from the cervicothoracic junction and extending the head and neck. The operation was performed under local anaesthesia with the patient awake and in a sitting position, which overcame

the difficult problem of intubation for general anaesthesia. It also allowed continuous intraoperative monitoring of nerve function in the limbs. Correction was achieved gradually over some days in an articulated plaster jacket incorporating the head and neck. In 1962 Law² described ten patients with ankylosing spondylitis treated by a cervical osteotomy under general anaesthesia, with one death. Since then there have been few reports of this hazardous procedure in more than a few patients. The only large series is by Simmons³ who operated on 42 patients under local anaesthesia using a technique similar to that of Urist,¹ except that the deformity was corrected intraoperatively by manual extension of the neck before immobilising the patient in a halo-jacket.

This paper describes the technique and results of cervical osteotomy in 15 patients with ankylosing spondylitis.

PATIENTS AND METHODS

Between 1986 and 1994, 15 patients with ankylosing spondylitis who had developed severe flexion deformities at the cervicothoracic junction were treated by an extension osteotomy of the cervical spine at the Princess Margaret Rose Orthopaedic Hospital, Edinburgh. The operations were carried out by the author with the patient in the prone position under general anaesthesia and wearing a halo-jacket. There were 13 men and 2 women whose mean age at operation was 48 years (35 to 67). The mean duration of symptoms before surgery was 16 years (9 to 35) and the indication was the inability of the patient to see straight ahead. One patient had a severe chin-on-chest deformity which interfered with eating and had caused marked weight loss. None had neurological abnormalities. In three patients the flexion deformity had deteriorated rapidly over a period of up to six months before osteotomy. All three were found to have had a fracture at the C5/6 level which had not been recognised; two had developed a pseudarthrosis and one had bony union in a deformed position.

Three patients who also had flexion deformity of the hip had been treated by soft-tissue release and total hip replacement two to three years before the cervical osteotomy. Two other patients also had severe flexion deformity at the thoracolumbar junction which had been treated by extension osteotomy of the lumbar spine at 4 and 15 months

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before the cervical osteotomy. The lumbar osteotomy had extended the spines by 35° and 40° respectively, but this had not been enough to allow the patient to see straight ahead.

Three patients had cardiac problems; two had had coronary thromboses and one an aortic valve replacement.

Preoperative management. The patients were admitted to hospital several days before operation and fitted with a lightweight underarm plaster jacket. A halo was applied under local anaesthesia and connected to the jacket by means of a single upright support on either side (see Fig. 4). No attempt was made to correct the deformity at this stage, and several days were allowed for adaptation to the halo-jacket and any necessary modifications. Patients with large obese abdomens were not accepted for this operation since this would interfere with the secure fitting of the halo-jacket and stabilisation of the neck until the osteotomy had healed.

The supports for the halo were hinged at the level of the body of C7 on both sides. These hinges allowed extension of the neck during the operation, but were locked at all other times. The upright supports were designed to allow adjustment in height as the neck was extended.

Technique of operation. General anaesthesia was induced while the patient was wearing the halo-jacket and an endotracheal tube passed, using a fibre-optic laryngoscope. The patient was then turned prone onto the operating table, which had slight head-up tilt to accommodate the flexion deformity at the cervicothoracic junction, and lifted clear of the table by large foam blocks placed beneath the chest and pelvis. The shoulders were often stiff, requiring placement of the arms by the sides.

The cervicothoracic spine was exposed from C6 to T1 through a midline longitudinal incision. The site of the osteotomy at the C7/T1 level was identified by locating the most caudal bifid spinous process, which was usually at C6, or from a lateral radiograph taken on the table.

The spinous processes of C6 and C7 were excised. The ligamentum flavum was not usually completely ossified in the midline of the interlaminar space, and it was possible to incise or nibble this layer with fine bone rongeurs to expose a small area of dura. This was often adherent to the under surface of the ossified structures, and required careful separation. Complete laminectomy was performed at C7 with removal of the inferior portion of the lamina of C6 and the superior portion of the lamina of T1. The eighth cervical nerve roots were completely exposed to beyond their intervertebral foramina by removal of the fused posterior facet joints. The pedicles of C6 and T1, lying above and below the C8 nerve roots, were then nibbled away to avoid pinching as the osteotomy closed. The adjacent lateral margins of the osteotomy slots were in the transverse plane and between 1 and 1.5 cm wide depending on the degree of correction required (Figs 1 and 2).

Spinal cord monitoring was performed throughout the procedure using somatosensory evoked potentials and an epidural electrode placed above the osteotomy. If there was

any concern, a wake-up test was performed, and the patient asked to move the lower limbs.⁴

To close the osteotomy, the hinges on the halo were unlocked. The surgeon then gripped the halo and extended the head and neck while directly viewing the exposed dura. This should become wrinkled during extension, showing that the spinal cord is being relaxed rather than stretched. The ankylosed anterior column of the cervical spine often fractured with a snap, after which it was relatively easy to place the head in a corrected position and lock the halo.

In the three most recent patients (cases 13 to 15), the osteotomy was stabilised additionally using internal fixation. A Luque rectangle with 3/16 inch rods was bent to conform to the corrected spine and secured by Drummond buttons and wires passed through the bases of the spinous processes at two levels above and below the osteotomy site.⁵ Sublaminar wires were not used because of ankylosis of the posterior bony elements and the possibility of tearing the underlying adherent dura.

The bone removed while performing the osteotomy was cut into strips and placed on either side of the spine across the osteotomy site, but not in the midline to avoid impingement on the exposed dura. Graft was also placed across the

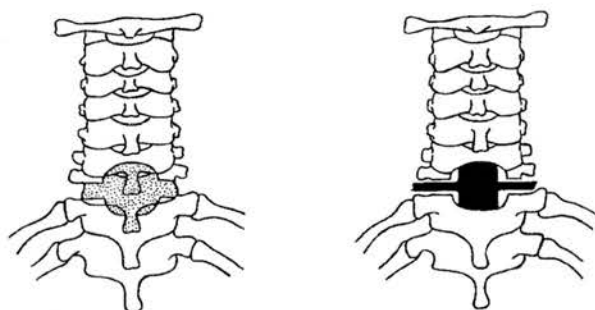


Fig. 1

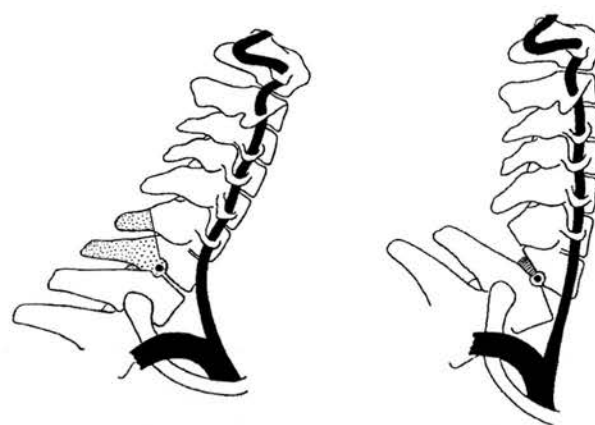


Fig. 2

Figures 1 and 2 - Diagrams to show the method of extension of the cervical spine at the C7/T1 level.

Table I. Details of 15 patients with ankylosing spondylitis treated by extension osteotomy of the cervical spine. The level of osteotomy was at C7/T1 in all

Case	Age (yr)	Sex	Previous surgery (spine/hips)	Preoperative kyphosis (degrees)	Postoperative lordosis (degrees)	Total correction (degrees)	Loss of correction (degrees)	Complications
1	59	M	Bilateral THR* at 52 years	20	50	70	20	Unilateral arm pain - resolved Haematemesis
2	55	M		Pseudarthrosis C5/C6 23	29	52	10	
3	38	M		20	33	56	8	Subluxation C7/T1 - 10% Unilateral arm pain - resolved
4	67	F		30	15	45	4	
5	43	M		10	40	50	0	Dysphagia - resolved
6	47	M		45	5	50	0	Subluxation C7/T1 - 40% Unilateral C8 palsy - resolved Dysphagia - resolved
7	38	M	THR at 35 years	Healed fracture C6 22	30	52	20	
8	45	M		20	48	68		Quadruparesis - no recovery
9	52	M		25	30	55	5	
10	46	M		20	40	60	0	
11	35	M	Lumbar osteotomy L2/L3 at 35 years	10	40	50	0	
12	40	M		24	6	30	0	Subluxation C7/T1 - 50% Unilateral arm pain - resolved Pseudarthrosis C7/T1 - anterior fusion
13	38	F		15	41	56	5	Unilateral C8 palsy - partial recovery
14	50	M	Bilateral THR at 47 years	30	20	50	10	Subluxation C7/T1 - 50% Unilateral arm pain - resolved Deep infection Pseudarthrosis C7/T1 - anterior fusion
15	51	M	Lumbar osteotomy L2/L3 at 49 years	35	36	71	3	Dysphagia - resolved

* total hip replacement

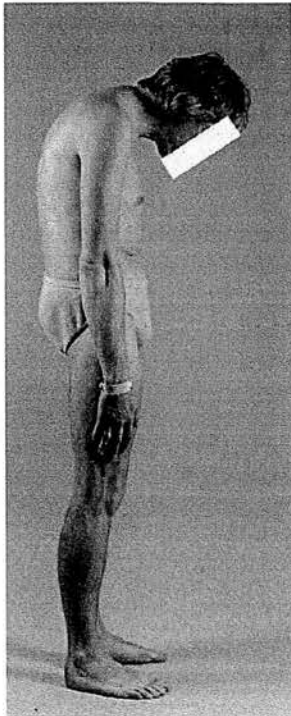


Fig. 3



Fig. 4



Fig. 5

Case 11. Figure 3 - A 35-year-old man with a severely flexed posture and a cervicothoracic kyphosis. Figure 4 - Four months after extension osteotomy of the lumbar spine, the flexed posture has been partially corrected but he is still unable to see straight ahead. A halo-jacket has been applied before a cervical osteotomy. Figure 5 - One year after a cervical osteotomy, he is fully corrected.

sites of the unhealed cervical fractures which were present in two patients.

Postoperative care. The patient was encouraged to take a few steps the day after operation. The position of the head and the ability to look straight ahead were assessed. If the head was not in the correct position and internal fixation had not been used the hinges were released and slight adjustments made under adequate analgesia. The patient usually left hospital ten days after operation and remained in the halo-jacket for three months. Radiographs taken at this time were difficult to assess and it was often necessary to use lateral tomography. When the osteotomy had healed, the halo-jacket was removed and a firm collar applied

which was worn for a further three months to allow consolidation.

The postoperative correction of the spine was measured on lateral radiographs using the Cobb method from the vertebra above to the vertebra below the site of osteotomy.

RESULTS

The osteotomy was performed at the C7/T1 level in all patients (Table I). Twelve had no internal fixation (Figs 3 to 7) and three had a Luque rectangle secured to the posterior spine by Drummond wires (Figs 8 and 9).

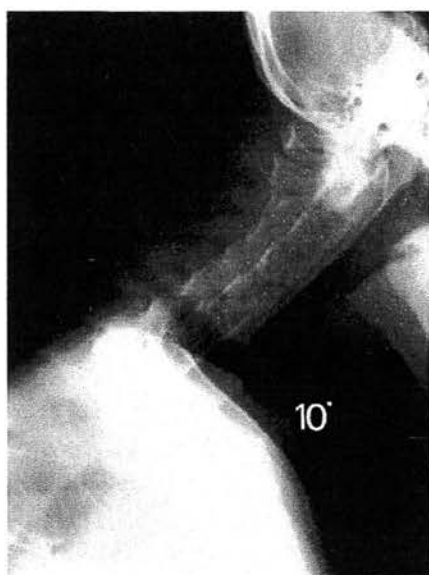


Fig. 6



Fig. 7

Figure 6 – Preoperative lateral radiograph. Figure 7 – Radiograph one year after cervical osteotomy. The osteotomy is soundly healed and the neck has been extended 50°.

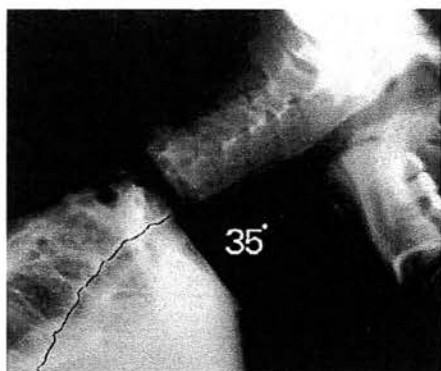


Fig. 8

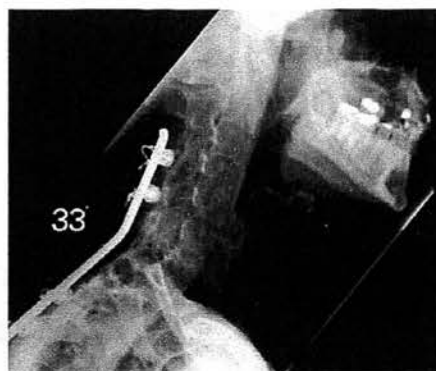


Fig. 9

Case 15. Figure 8 – Lateral radiograph of a 51-year-old man with a cervicothoracic kyphosis of 35°. Figure 9 – One year and four months after cervical extension osteotomy at the C7/T1 level with internal fixation using a Luque rectangle with spinous process wiring. The neck has been extended through 68° and the osteotomy is soundly healed.

Before surgery, there was a mean kyphosis of 23° (10° to 45°) at the cervicothoracic junction. Lateral radiographs of the spine after the operation showed that closure of the posterior wedge had fractured the ossified anterior longitudinal ligament and opened the anterior disc space at the C7/T1 level, producing a mean cervical lordosis of 31° (5° to 50°). The mean total correction was 54° (30° to 71°); all of the patients were able to see straight ahead. Follow-up was for a mean of 18 months (9 months to 3 years) and the mean loss of correction at the osteotomy site during this period was 6° (0° to 20°). Five patients had no loss of correction. The greatest loss was 20° during the first six months in two patients (cases 1 and 7) treated without internal fixation. After six months the fusion at the osteotomy became fully consolidated and there was no further loss of correction after two years and 18 months, respectively.

The unhealed fractures which were found in two patients (cases 1 and 12) before operation united during immobilisation in a halo-jacket.

One patient (case 13), whose cervical correction was well maintained at 51° , developed an increasing thoracolumbar kyphosis and may require an extension osteotomy of the lumbar spine at a later stage.

None of the patients had been able to work before operation but afterwards four were able to obtain employment six to nine months after osteotomy.

Complications

Neurological. After operation all the patients became independently mobile in their halo-jackets but one man (case 8) whose cervical kyphosis had been corrected by 68° ,

became quadraparetic one week later. There was no apparent reason: he had been walking normally and check radiographs showed no obvious displacement at the osteotomy site. The head and neck were allowed to flex to their preoperative position but there was no recovery.

Two patients developed unilateral compression of one C8 nerve root at the site of the osteotomy, causing some weakness and hypoaesthesia in the hand. This was noted one to two days after operation and was treated by distracting the halo on the jacket. One patient (case 6) recovered to normal over a period of two months; the other (case 13) recovered full motor function but was left with mild hypoaesthesia affecting the little and ring fingers.

Four patients had intermittent pain in one arm in the C8 dermatome but no neurological deficit. No treatment was given and the pain resolved completely at from a few days to two weeks.

Subluxation. There was subluxation with forward displacement of the seventh cervical vertebra on the first thoracic at the site of the osteotomy in four patients. This was not seen to happen at operation and was first noted on the immediate postoperative radiographs (case 12; Figs 10 and 11). The halo was distracted on the jacket but with very little improvement. One patient (case 3) with 10% forward displacement at the osteotomy healed satisfactorily with 8% loss of correction. A second patient (case 6), with 40% forward displacement at the osteotomy, had unilateral signs of compression of the C8 nerve root which fully recovered spontaneously within two months; the osteotomy healed without loss of correction. The other two patients (cases 12



Fig. 10

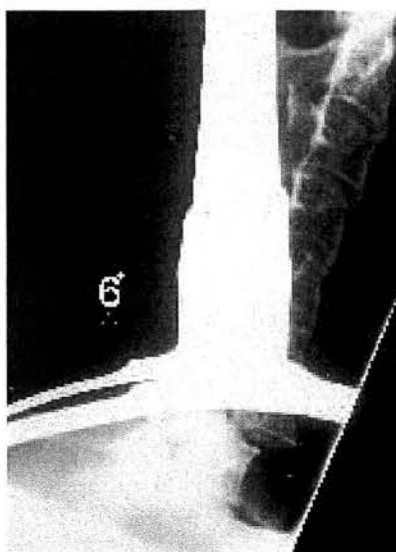


Fig. 11



Fig. 12

Case 12. Figures 10 and 11 – Pre- and postoperative lateral radiographs showing 50% forward subluxation of C7 on T1 at the site of the osteotomy. Figure 12 – A pseudarthrosis at the osteotomy site was treated by an anterior spine fusion. Nine months after the initial operation, there is healing with no loss of correction.

and 14) had 50% forward displacement, and unilateral arm pain in the C8 distribution but no neurological deficit. Both developed nonunion which required anterior cervical fusion using an inlay autogenous iliac bone graft at four and six months after the osteotomy. One of these patients gained solid fusion (Fig. 12); the other did not heal and was found to have a deep infection.

Dysphagia. Three patients had difficulty in swallowing because of retropharyngeal swelling at the site of the osteotomy, which resolved completely over ten days. Three patients with a severe chin-on-chest deformity found that after correction their mouths tended to hang open, but this resolved in a few weeks.

Haematemesis. One week after operation one patient (case 1) had a haematemesis which settled with conservative treatment.

DISCUSSION

The indication for extension osteotomy of the cervical spine in patients with ankylosing spondylitis is severe flexion deformity at the cervicothoracic junction which prevents the patient from seeing straight ahead and limits the field of vision to the area around the feet.

It is important to assess the contribution of all levels of the spine and hips to the overall flexed posture. A thoracolumbar kyphosis is the most common deformity and is best treated by an extension osteotomy in the lumbar region to create a compensatory lordosis. Severe flexion deformities at the hips can often be overcome by soft-tissue release and total hip replacement. Less frequently, the major deformity is at the cervicothoracic junction and this requires an extension osteotomy of the cervical spine. When other regions are affected, it is best to start with surgery on the hip, then perform lumbar osteotomy and finally a cervical osteotomy if this is still required. An attempt to compensate fully for a severe flexion deformity of the cervical spine by means of a lumbar osteotomy alone may unbalance the patient and even then fail to allow vision straight ahead. Three of the patients now reported had prior total hip replacements and two had osteotomies of the lumbar spine (Figs 3 to 7).

The preferred site for the osteotomy is between the C7 and T1 vertebrae. At this level, the vertebral arteries pass in front of the transverse processes of C7 before passing into and through the foramen transversum of C6 and are less liable to kink when the neck is extended (see Fig. 2). The spinal canal is also larger in this region.

Closure of the posterior wedge excision is performed manually and a halo-jacket is required to immobilise the neck in the corrected position until the osteotomy becomes stable. A preliminary anterior osteotomy⁶ is unnecessary because the ankylosed anterior column fractures easily. It can be difficult or impossible to approach this area anteriorly with the patient uncorrected and the chin flexed onto the chest. Correction at the site of the osteotomy occurs by

rupture of the ossified anterior longitudinal ligament and the opening of the C7/T1 disc space anteriorly. Care is needed to avoid overcorrection.

A cervical osteotomy is much more hazardous than lumbar osteotomy in patients with ankylosing spondylitis. Three of the 15 patients reported developed neurological deficits and one became quadriparetic. There were no neurological complications in a personal series of 50 extension osteotomies of the lumbar spine in patients with ankylosing spondylitis.⁷ Neurological complications after cervical osteotomy have several possible causes.

First, the neural structures may be stretched if the osteotomy has been cut incorrectly and the axis of angulation in the sagittal plane is at or posterior to the spinal cord. This would result in immediate injury to the cord when the neck is extended. None of the patients in this series had this complication: the osteotomy was cut far laterally (Fig. 1) to prevent impingement of the posterolateral bony masses as the neck is extended. Wrinkling of the dura during extension indicates that the neural structures are being relaxed rather than stretched.

Secondly, the C8 nerve roots may be compressed in their intervertebral foramen if sufficient bone has not been removed from the pedicles above and below the osteotomy. Two patients were affected by this, but both spontaneously recovered full motor function; one had mild residual hypoaesthesia. Four patients complained of transient unilateral C8 root pain, but without neurological deficit, which settled quickly.

Thirdly, neurological complications may be due to instability and subluxation at the osteotomy. Subluxation with forward displacement of C7 on T1 occurred in four patients. This was the main technical problem which occurred when the neck was being extended manually and was due to disruption of the posterior longitudinal ligament, the hinge about which angulation takes place (Fig. 11). Subluxation was not apparent during the operation, possibly being disguised by the wide bony decompression and angulation at the osteotomy site. This complication was first described by Herbert⁸ and was the cause of death in one of his three patients undergoing cervical osteotomy. To prevent cord compression due to subluxation it is important to perform an extensive mid-line bony decompression above the osteotomy level so that the lamina of C6 does not impinge on the spinal cord if the vertebral body becomes displaced forwards. A halo-jacket does not provide complete postoperative stability and 'snaking' of the cervical spine can occur in the sagittal plane if there is no internal fixation, resulting in transient displacement. This may have caused the quadripareisis in the patient (case 8) who had no internal fixation and was mobilising in his halo-jacket one week after operation. The addition of internal fixation may prevent loss of correction and displacement postoperatively but cannot prevent subluxation during manual extension of the neck (Fig. 11).

If subluxation occurs, there is a high incidence of nonunion at the osteotomy site, especially with displacement. Both patients with 50% subluxation developed nonunion which required anterior spine fusion.

Cervical osteotomy in ankylosing spondylitis is a difficult and potentially hazardous procedure but a successful

operation allows the patient to see straight ahead and provides major functional and psychological benefits.

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PRACTICE OF SURGERY

Operative Spinal Surgery

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Surgery for ankylosing spondylitis

Michael J. McMaster

INTRODUCTION

Ankylosing spondylitis is a disease which usually occurs in young men and is characterized by a chronic inflammatory process which first affects the sacroiliac joints and then extends to the lumbar, thoracic and often the cervical regions of the spine. On occasion the larger joints of the extremities are also affected. The inflammation occurs at the insertion of the joint capsules and ligaments into bone and as the process settles these structures slowly ossify resulting in ankylosis of the spine. After a number of years the disease 'burns itself out' but before this there is a tendency for the spine to develop an increasing flexion deformity.

Fortunately the majority of patients are only mildly to moderately affected and do not become severely deformed. However, a few are much more severely affected and by the fourth decade have progressed to severe or total spinal ankylosis in an extremely deformed position. At this stage, ankylosing spondylitis can be one of the most crippling conditions seen by the orthopaedic surgeon.

The characteristic spinal deformity in these severely affected patients is a flattening of the normal lumbar lordosis, and an increased smooth thoracic kyphosis with the head and neck thrust forwards. Occasionally there is also a flexion deformity at the cervicothoracic junction. The entire vertebral column from sacrum to occiput may be ankylosed by bone in this deformed position and the patient is bent forwards and forced to look at the ground. To see in front of him, the patient attempts to hyperextend his hips, but unfortunately this may not be possible because the hips are also frequently involved and may have flexion contractures. The patient, therefore, flexes his knees and adopts the crouched stance characteristic of ankylosing spondylitis (Fig.

19.15). This ugly posture is not only functionally disabling but is also psychologically disturbing and the patient may confine himself to the house because of his appearance.

INDICATIONS FOR SURGICAL CORRECTION

The main indication for the surgical correction of the severely flexed posture in a patient with ankylosing spondylitis is the patient's inability to see straight ahead with the field of vision limited to the area around his feet.

ASSESSMENT OF THE DEFORMITY

Before proceeding to surgery it is important to assess the contribution of all levels of the spine from the occiput to the sacrum as well as the hips to the flexed posture. Frequently, all of these regions are affected but some are more deformed than others.

Severe flexion contractures of the hips can often be corrected by total hip replacements and this may be sufficient by itself to allow the patient to see straight ahead (Fig. 19.1).

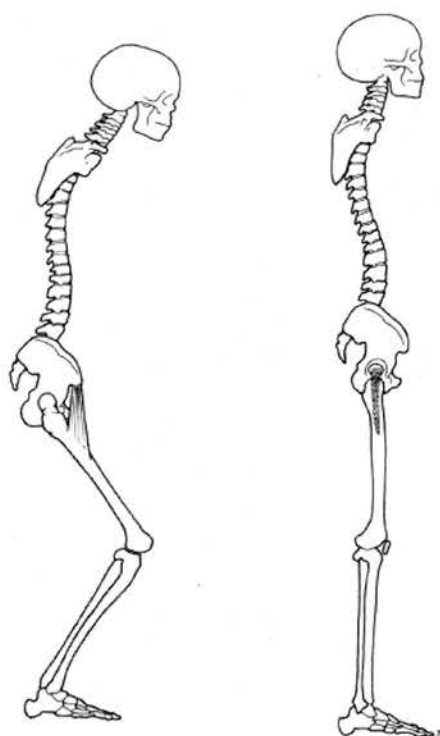


Fig. 19.1

If there are significant hip flexion contractures, the overall flexed posture can often be improved by total hip replacements and soft tissue release.

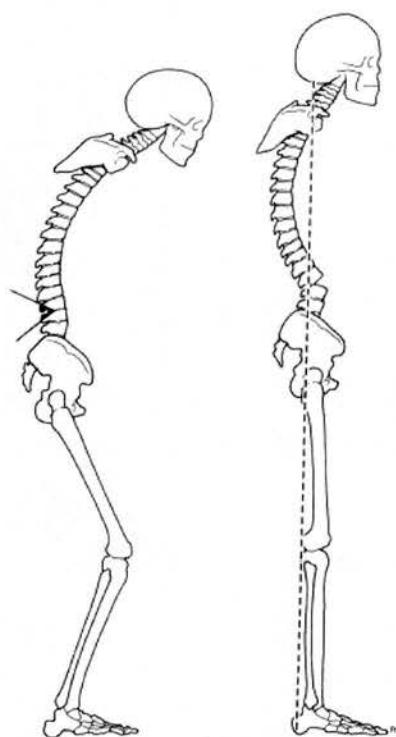


Fig. 19.2

A flexion deformity mainly in the thoracolumbar spine is overcome by creating a compensatory lumbar lordosis.

A spinal osteotomy is only indicated if the hips are not significantly deformed, or if after hip operations the patient is still unable to see straight ahead. The thoracic spine is usually the most flexed region but an osteotomy at this level does not help because the ankylosed thoracic cage prevents extension of the spine. A thoracic deformity is best overcome by the creation of a compensatory lumbar lordosis by means of a lumbar osteotomy (Fig. 19.2). Ideally the lumbar spine should be extended until the patient can see straight ahead. The centre of gravity of the upper body should then lie just behind the site of the osteotomy helping to maintain the correction.

In a few patients, the major flexion deformity is at the cervicothoracic junction. Correction of this deformity by a lumbar osteotomy could possibly unbalance the patient and still not enable him to see straight ahead (Fig. 19.3a). These patients require an extension osteotomy of the cervical spine (Fig. 19.3b).

On occasion, the decision whether to perform a lumbar or cervical spinal osteotomy is not always clear, and in these circumstances one should favour a lumbar osteotomy as it is less hazardous for the patient.

ADVANTAGES OF SURGERY

The surgical correction of the severe flexed posture in these patients provides major functional and psychological benefits. It not only allows the patients to see straight ahead but also improves their self-esteem and often allows them to return to work. In addition, it relieves the compression of the abdominal contents and improves the capacity for diaphragmatic breathing which is essential in these patients who have an ankylosed thoracic cage.

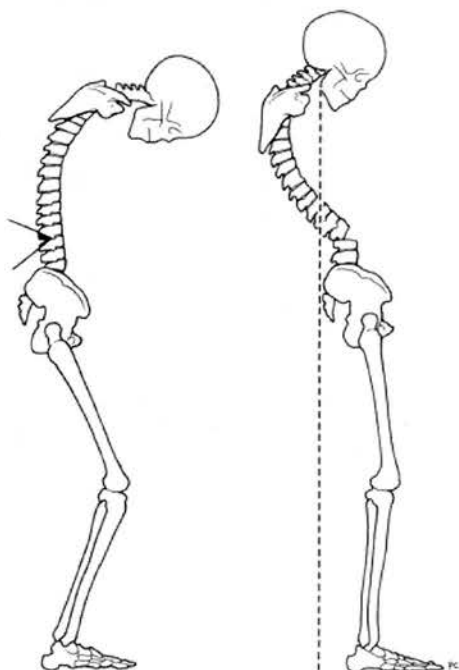


Fig. 19.3a

If the major flexion deformity is at the cervicothoracic junction, an attempt to correct this by means of an extension osteotomy in the lumbar spine may unbalance the patient. This patient requires a cervical osteotomy.

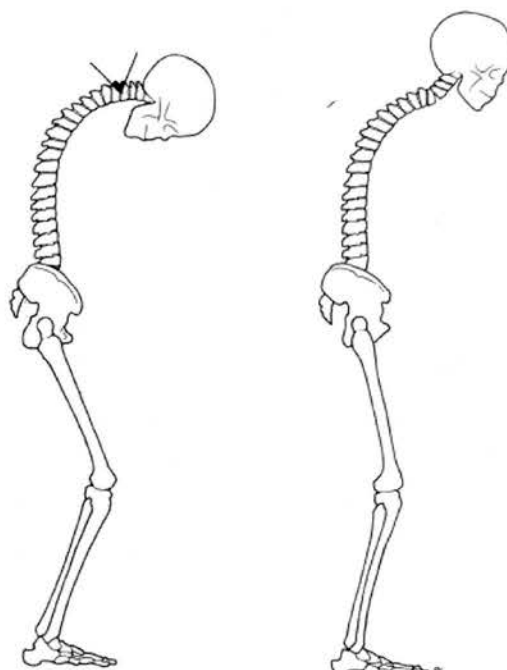


Fig. 19.3b

A flexion deformity occurring mainly at the cervicothoracic junction is overcome by a cervical osteotomy.

LUMBAR OSTEOTOMY

Smith-Petersen, Larsen and Aufranc recognized the plight of these unfortunate patients with ankylosing spondylitis and in 1945 introduced the operation of lumbar spinal osteotomy, in which the spine is hyperextended in the lumbar region enabling the patient to see straight ahead. They obtained successful results in six patients, all of whom were operated upon by a posterior approach and the lumbar spine was osteotomized at one, two or three levels. These osteotomies were confined to the laminae and articular facets and did not involve the vertebral bodies. Correction was obtained by forced manipulation and hyperextension on the operating table after which a plaster jacket was applied and maintained for 2 months followed by a brace for 1 year.

La Chapelle (1946) and Herbert (1948) believed that a posterior osteotomy alone was not always sufficient to give good correction especially if there was considerable ossification of the anterior longitudinal ligament. They used a two-stage anterior and posterior approach which enabled them to divide all of the ossified ligaments under direct vision. Most other surgeons, however, found that a single level posterior osteotomy was sufficient and multiple level osteotomies were not necessary as all of the correction occurred at one level.

Since this time only a relatively few surgeons have attempted this major procedure and many have reported a high incidence of serious complications (Adams 1952, Herbert 1959, Law 1959, McMaster 1962, Goel 1968, McMaster & Coventry 1973, Simmons 1977, Thomasen 1985). Bradford et al (1987) reviewed the complication rate in those series with five or more cases and found that in these 250 patients, there was a 7% mortality and a 6% incidence of neurological complications. However, McMaster (1985) has shown that the

majority of these problems can be overcome by:

- a. Careful attention to the technique of performing the osteotomy so that the axis of correction lies anterior to the spinal cord
- b. Adequate decompression of the spinal cord and nerve roots
- c. The use of instrumentation to provide a slow and finely controlled closure of the osteotomy and rigid internal fixation.

The following operative description is based on these principles.

Operative technique

Endotracheal anaesthesia is necessary as it is essential to maintain a good airway during the surgery. However, intubation can be difficult due to the rigid flexion deformity in the cervical spine and a fibre-optic laryngoscope, over which the endotracheal tube is passed, is often necessary if tracheostomy is to be avoided.

Once anaesthetized, the patient is turned onto the operating table, with care to avoid injury to the ankylosed neck, and positioned face down with the chest and pelvis supported on firm foam blocks which leave the abdomen hanging free. This position reduces the intra-abdominal pressure and when combined with hypotensive anaesthesia and infiltration of the operation site with a 1:400 000 solution of adrenaline, greatly reduces bleeding during the operation and makes it easier to identify the neural structures (Malcolm-Smith & McMaster 1983). Careful positioning on the operating table is important but can be difficult due to the severe flexed posture of the spine. The flexion/extension hinge on the operating table should be at the level of the hips because this is later used to help extend the patient.

The preferred site for the spinal osteotomy is either between the second and third lumbar vertebra or between the third and fourth. These levels are in a region where there is a relatively spacious spinal canal containing the cauda equina which is less easily injured than the spinal cord and are also far enough from the sacrum to allow the application of an internal fixation device. The severity of the ankylosis as seen on the spinal radiographs is not important in deciding on the site of the osteotomy.

The lumbar spine is exposed through a midline longitudinal incision extending from L1 to L5. The paraspinal muscles are stripped subperiosteally from either side of the spinous processes and laminae exposing the spine from two levels above to two levels below the proposed site of the osteotomy. Identification of the site of the osteotomy is made by noting the relative position of the last rib or by counting the vertebrae up from the sacrum. However, the anatomical details are often obscured by ossification of the ligaments, and an intra-operative radiograph may be necessary to help locate the exact level. At the site of the osteotomy, the muscles are stripped laterally out to the tips of the transverse processes and the operative field held exposed by deep self-retaining retractors.

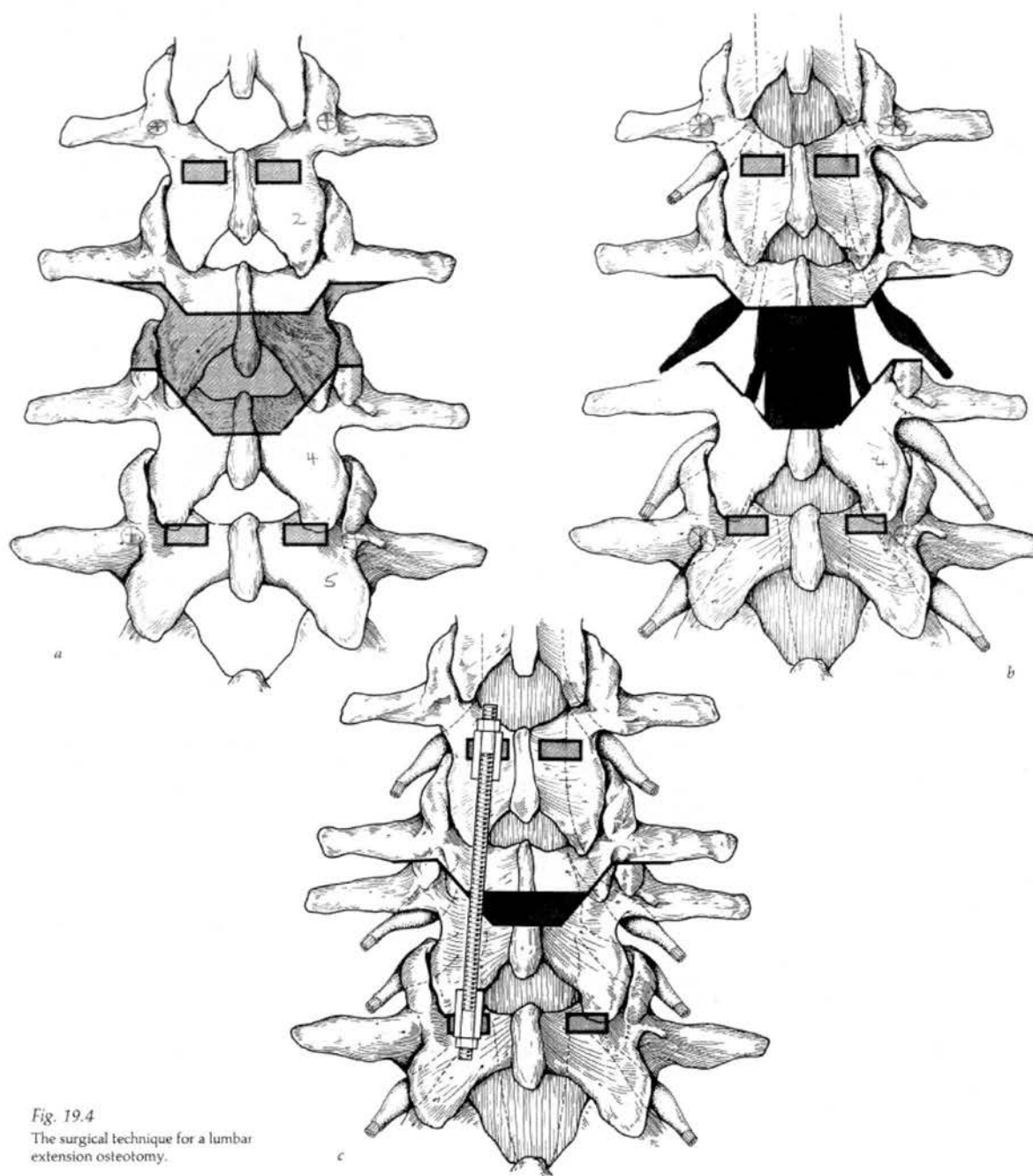


Fig. 19.4
The surgical technique for a lumbar extension osteotomy.

The bony sites for the application of the internal fixation device must be prepared before cutting the osteotomy. Using a 10 mm wide osteotome, notches are cut in the ossified ligamentum flavum and medial parts of the fused interfacetal joints on either side of the spinous processes of the vertebrae above and below the site of the osteotomy (Fig. 19.4a). Initially these notches are made only in the outer cortex of the bone and are completed by hammering a sharp Harrington hook (No. 1251), on an introducer, until the hook is firmly and deeply embedded within the bone. The hook does not necessarily enter the spinal canal because the bone at the site of the insertion has been greatly thickened by the disease process. The upper two hooks should lie over the top of the lamina of the vertebra at the level above the osteotomy site, and the lower two hooks around the bottom of the lamina at the level below the osteotomy site. Once each bony site is prepared for later use, the sharp hook is removed. Hammering on the hooks after the osteotomy has been cut is dangerous because it could displace the vertebra and cause neural damage.

The osteotomy is begun using heavy bone cutters to remove a posteriorly based wedge of bone which includes the adjacent spinous processes and the ossified interspinous ligament and exposes the interlaminar space. The ossified ligamentum flavum in the midline is nibbled away with rongeurs until a small area of dura is exposed. A midline approach is best because the ligamentum flavum may not be completely ossified in this region and there is also the greatest potential space between the ligamentum and the underlying dura. The dura is often atrophic and adherent to the under surface of the ossified structures and can easily be torn unless it is carefully separated using a fine blunt dissector. Once separated, the dura is widely exposed in the interlaminar space by using upcutting laminectomy rongeurs to remove the overlying bone. The fused interfacetal joints at the same level are identified and the osteotomy extended from the midline across these joints at an angle of 45° on either side (Fig. 19.4b). The bone may be up to 2 cm thick in this region

and the osteotomy is at first only deepened through the outer cortex into the cancellous bone using osteotomes and gauges directed laterally away from the dura. Finally, small upcutting laminectomy rongeurs are used to advance the slot anteriorly through the inner cortex and into the intervertebral foramen on either side exposing the nerve roots. The bony sides of the osteotomy slot should be parallel and up to 18 mm wide depending on the degree of spinal correction that is required. Symmetrical amounts of bone should be removed from either side of the spine. It is very important to ensure that the transverse axis of eventual closure of the osteotomy, about which correction will occur, lies anterior to the cauda equina. This means that the osteotomy must be deepened anteriorly so that its apex lies at the anterior margin of the spinal canal (Fig. 19.5a, b), and is also extended laterally on either side to expose the nerve roots outside their intervertebral foramina (Fig. 19.4b).

If the osteotomy is cut correctly, the cauda equina will be relaxed as the spinal column is extended and the osteotomy closes. An incorrectly cut osteotomy will allow hinging on the bony structures posterior to the neural elements and result in serious neurological complications due to stretching of the cauda equina. It is also important to remove enough bone from the pedicles above and below each intervertebral foramen to ensure that the nerve roots are not pinched as the osteotomy is closed (Fig. 19.5a, b). This is done using fine osteotomes and rongeurs while protecting the nerve roots with a blunt dissector.

In the past, the osteotomy has been closed using the method described by Smith-Peterson et al (1945) in which the surgeon applies direct manual pressure to the bone on either side of the

osteotomy site while unscrubbed assistants extend the upper body and thighs. The ossified anterior vertebral column is fractured by this pressure and this often occurs with a sudden snap making it difficult to prevent displacement and overcorrection which could result in serious neurological complications (McMaster & Coventry 1973). Even if there are no immediate neurological complications, the spine is now very unstable and therefore potentially dangerous and requires to be controlled by the application of plaster shells before the patient is removed from the operating table.

In the technique described here, these problems are overcome by the use of instrumentation which is applied directly to the bony spine to close and stabilize the osteotomy. Once the osteotomy has been completed, no

attempt should be made to close the wedge until the internal fixation device has been applied. It is possible although difficult in the confined space to thread a compression rod through the normal Harrington hooks once they are seated in their prepared sites above and below the osteotomy. In order to overcome this problem, blunt Harrington hooks (No. 1253) were modified in the Bio Engineering Laboratories at the Princess Margaret Rose Hospital and a slot was cut in the top of the hook to enable it to accept a compression rod after the hook had been placed in the bone. Similar hooks are available commercially, but these are too slim and the shoe can cut through the relatively soft bone found in these patients. The Harrington No. 1253 hook has a much broader shoe and is therefore less likely to cut through the bone when compression is applied.

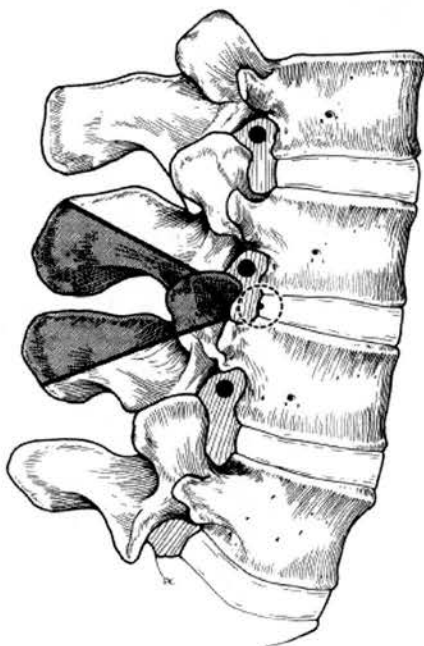


Fig. 19.5a

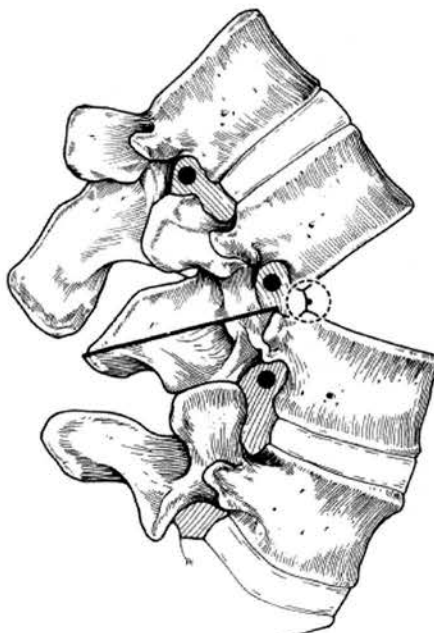


Fig. 19.5b

The surgical technique for a lumbar spine extension osteotomy.

Figs 19.6 and 19.7

A man with a severe flexion deformity in the thoracolumbar region.



Fig. 19.7

Figs 19.8–19.10

Correction has been achieved by a lumbar spine extension osteotomy performed at the level between the second and third lumbar vertebrae. The ossified anterior longitudinal ligament has fractured and there is an opening of the anterior disc space. One year later the osteotomy is fused and the patient can see straight ahead.



Fig. 19.8



Fig. 19.9



Fig. 19.10

Four of these modified hooks are inserted by means of an introducer into their prepared fixation sites above and below the osteotomy and gently tapped into position. A Harrington compression rod is pressed down into the hooks on each side of the spine. These rods are held in place by special bushes which are advanced along the rod towards the osteotomy and into the base of each hook where they are enclosed by the hook but cannot pass through it. The osteotomy is closed by slowly tightening the nuts on the threaded rods, alternating from side to

side of the spine, and thus applying even compression on all four hooks. Hook holders are used to control the four hooks and prevent them from rotating out of position as the osteotomy closes and their relative position changes. As the osteotomy closes, the operating table is slowly extended at hip level and this partially relieves the pressure on the hooks and prevents them cutting out of the soft bone. The resistance to tightening the nuts gradually increases as the posterior wedge closes and then relaxes as the anterior vertebral column gives way.

This occurs either by a fracture of the ossified anterior longitudinal ligament with opening of the anterior disc space (Figs 19.8 and 19.9) or by a posterior wedge compression fracture with collapse of the posterior aspect of the vertebral body at the site of the osteotomy (Figs 19.11 and 19.12). It is possible, although not necessary, to encourage this second type of correction by decancellation of the posterior part of the vertebral body by means of a curette passed through the base of the pedicle (Thomasen 1985).



Fig. 19.11



Fig. 19.12

A lumbar spine extension osteotomy resulting in a posterior wedge compression fracture with collapse of the posterior aspect of the vertebral body at the site of the osteotomy.

The advantage of this technique is that it allows a slow and finely controlled closure of the osteotomy without any sudden displacement of the vertebrae. As the osteotomy closes, the dura which is well exposed in the midline is seen to wrinkle confirming that the neural elements are relaxed (Fig. 19.4c). If the dura does not wrinkle, the lateral parts of the osteotomy should be inspected to make sure there is no premature impingement of the bone posterior to the neural structures. The interlocking 'V' shape of the osteotomy makes it very stable and prevents any rotation. The compression rods prevent distraction and maintain correction.

Once the osteotomy is closed, a wake-up test is performed and the neurology in the lower limbs is checked. If there is a neurological deficit the fixation can be released. If all is well, bone chips removed during the osteotomy are placed posteriorly and also on both sides between the transverse processes of the vertebrae above and below the osteotomy. These transverse processes are much closer together once the osteotomy has been closed (Fig. 19.4c).

Complications

Gastric dilatation is a serious complication which may occur in the few days after lumbar spinal osteotomy. Extension of the spine may cause the third part of the duodenum to be pinched between the superior mesenteric vessels which pass over it and the spine which lies behind it. The obstruction usually resolves after a few days but if it is not recognized the stomach may become distended and the patient may vomit large quantities of fluid, with the danger of aspiration into the lungs. Because of the ankylosed thoracic cage, these patients cannot cough properly and easily develop aspiration pneumonia which can be fatal. To prevent this complication, a nasogastric tube is passed before the patient awakes from the anaesthetic and is retained in place for up to 48 h or until it is apparent that there is no obstruction.

Postoperative care

After the operation the patient is nursed on a firm bed and log rolled from side to back to side. External support is unnecessary because of the rigid internal fixation. Physiotherapy is required to encourage coughing and the removal of pulmonary secretions. The use of an absorbable subcuticular suture allows the application of a well-moulded underarm plaster jacket 5 days after the operation. This jacket is applied with the patient lying supine on the plaster table with a support under the osteotomy site. The patient is then allowed to walk and is usually fit to leave hospital 2 weeks after the operation. Light sedentary work is allowed after a few weeks but heavy lifting is forbidden until the spine is solidly fused. The plaster jacket is removed 9 months after the surgery provided that radiographs show a solid fusion. A Jewett brace is then worn during the day for a further 3 months and after this there are no restrictions (Figs 9.6 to 9.10).

Comment

Using this technique it is possible to perform a lumbar spinal osteotomy in patients with ankylosing spondylitis without the high incidence of major complications reported in other series (McMaster 1985). It is usually possible to extend the spine through 40° at the site of the osteotomy and I have used this technique on over 35 patients without any deaths or neurological complications.

However, ankylosing spondylitis is a progressive disorder, with exacerbations and remissions before the disease eventually 'burns itself out' and a lumbar spinal osteotomy is just one incident in this continuing process. McMaster & Coventry (1973) in a long-term review of the results of lumbar spinal osteotomy over 10 years found that once the osteotomy had fused there was usually no further loss of correction in this region but continuing active disease in the thoracic and cervical spine or hips could allow increasing flexion at these sites and detract from the overall initial correction. However, if the spinal deformity is already crippling, it is not possible to wait for the disease to become quiescent and correction lost at these sites can often be regained by either hip replacement or cervical osteotomy.

CERVICAL OSTEOTOMY

In some patients the major site of the spinal flexion deformity is at the cervicothoracic junction rather than in the thoracolumbar region. On occasion the cervicothoracic deformity may deteriorate rapidly and in these circumstances a stress fracture or pseudarthrosis may have developed in the ankylosed spine either spontaneously or following relatively minor trauma. In a few patients, the deformity may become so severe that the chin comes to rest on the chest and this not only prevents the patient from seeing straight ahead but also interferes with the ability to open the mouth and leads to severe weight loss.

The basic principles of a cervical osteotomy in a patient with ankylosing spondylitis are the same as for a lumbar osteotomy. However, the possible complication of tetraplegia is a much greater hazard. The major problem is that, unlike a lumbar osteotomy, it is not possible to rely on any effective form of internal fixation to help close the osteotomy or stabilize the spine post-operatively. Closure of the osteotomy is done manually and a halo body jacket is used to immobilize the neck in the corrected position until the osteotomy has healed and become stable.

A further problem is that the flexed position of the head and neck makes it extremely difficult if not impossible to pass an endotracheal tube to give a general anaesthetic. Urist (1958) and Simmons (1972, 1977) overcame this problem by performing the operation under local anaesthesia with the patient in a sitting position. This has the additional advantage of allowing continuous monitoring of the neurological status during the procedure but predisposes the patient to the risk of air embolism.

The surgical technique that I have used is very similar to that of Simmons (1977), except that the patient is operated upon under a general anaesthetic and in the prone position while wearing a halo body jacket. The problems of endotracheal intubation are overcome by the use of a fibre-optic laryngoscope.

Preoperative preparation

The patient is admitted to hospital several days before surgery and fitted with a lightweight underarm plaster jacket. A halo is then applied under local anaesthesia and connected to the plaster jacket by means of single upright supports on either side. No attempt is made at correction and the patient wears the halo jacket in the deformed position. Several days are allowed before surgery to enable the patient to adapt to the halo jacket and also to allow for any modifications that might be necessary.

The supports for the halo have hinges located at the level of the body of C7 on either side. These hinges allow the neck to be extended during the surgery but remain locked at all other times. It is also necessary for the upright supports to have a piston effect which can be unlocked and allow them to adjust in height as the neck is extended.

Operative technique

The patient is given a general anaesthetic while wearing his halo body jacket and an endotracheal tube passed using a fibre-optic laryngoscope. He is then turned prone onto the operating table and large foam blocks placed beneath the chest and pelvis. This lifts the patient off the operating table and accommodates for the flexion deformity at the cervicothoracic junction. The shoulders are often stiff and this necessitates placing the arms by the side.

The cervicothoracic spine from C6 to T1 is exposed through a midline longitudinal incision. The subcutaneous tissues and paraspinal muscles are infiltrated with a solution of 1:400 000 adrenaline and this helps control the bleeding. The preferred site for the osteotomy is between C7 and T1 (Figs 19.13a, 19.14a) and this level is located by noting the most caudal bifid spinous process which is usually at C6. If necessary the level can be confirmed by an intra-operative X-ray.

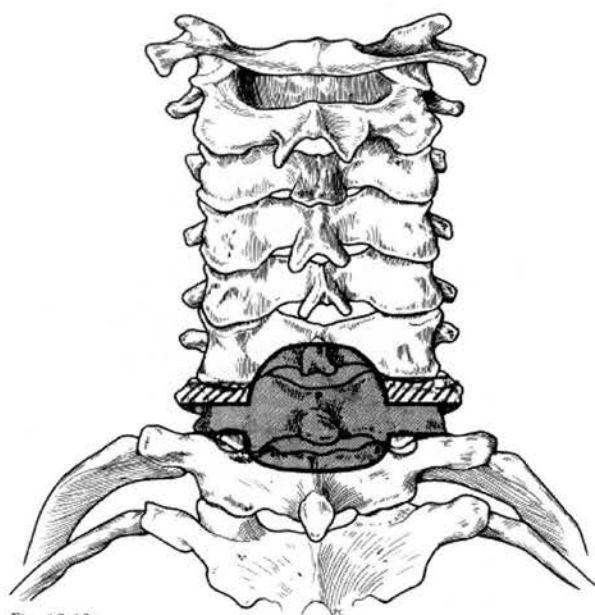


Fig. 19.13a
The surgical technique for a cervical spine extension osteotomy.

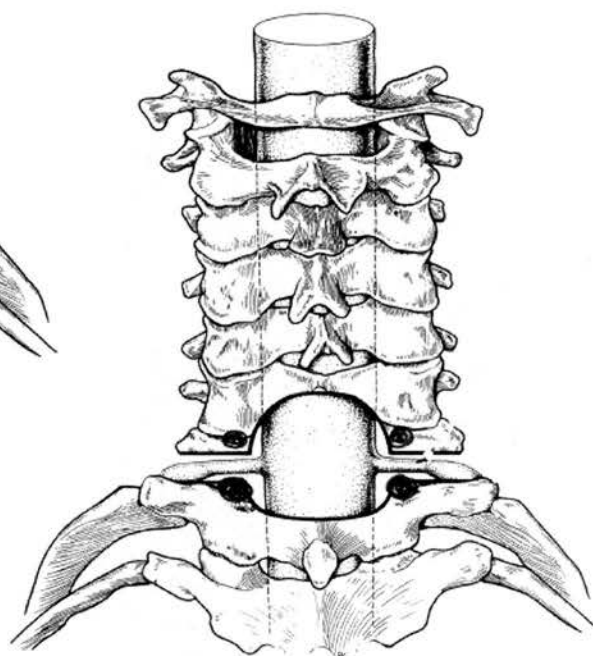


Fig. 19.13b

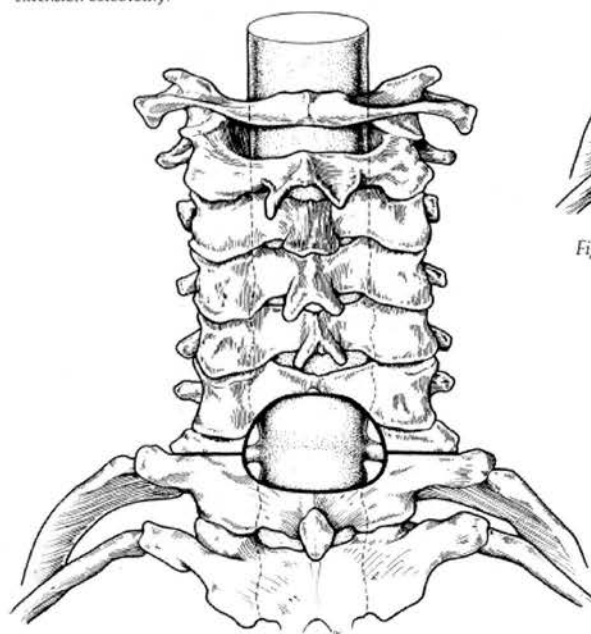


Fig. 19.13c

In this region the spinal canal is relatively spacious and the spinal cord and eighth cervical nerve roots are relatively more flexible. Any injury to the eighth cervical nerve roots will cause much less functional disability than injury to the nerve roots above this level. In addition the vertebral arteries pass into and through the foramen transversum of the vertebrae of C6 and above and are therefore much less liable to injury below this level (Fig. 19.14).

The spinous processes of C6 and C7 are excised using heavy bone cutters and it is not uncommon to find that the ligamentum flavum is not completely ossified in the midline in the interlaminar space. This allows the ligamentum to be incised or nibbled

away with fine bone rongeurs to expose a small area of dura. A midline approach is best because here there is the greatest potential space between the ligament and the underlying dura. However, the dura is often adherent to the under surface of the ossified structures and can be easily torn unless it is carefully separated with a fine curved dissector. Using small upcutting laminectomy rongeurs, a complete laminectomy is performed at C7 along with removal of the inferior portion of the lamina of C6 and the superior portion of the lamina of T1. This gives a wide exposure of the spinal cord and the eighth cervical nerve roots which pass at right angles into their intervertebral foramina. These nerve roots are completely exposed by removing the overlying bony structures

which are the fused posterior facet joints. The pedicles of C7 and T1 lying above and below the nerve roots are nibbled away to prevent a pinching effect when the osteotomy is closed. The adjacent margins of the osteotomy slots lie in the transverse plane and are between 1 and 1.5 cm wide depending on the degree of correction required (Fig. 19.13b).

It is most important to ensure that the transverse axis of eventual closure of the osteotomy lies anterior to the spinal cord. This means that the osteotomy must be deepened anteriorly so that its apex lies at the posterior longitudinal ligament and is also extended laterally on each side to expose the eighth cervical nerve roots outside their intervertebral foramina.

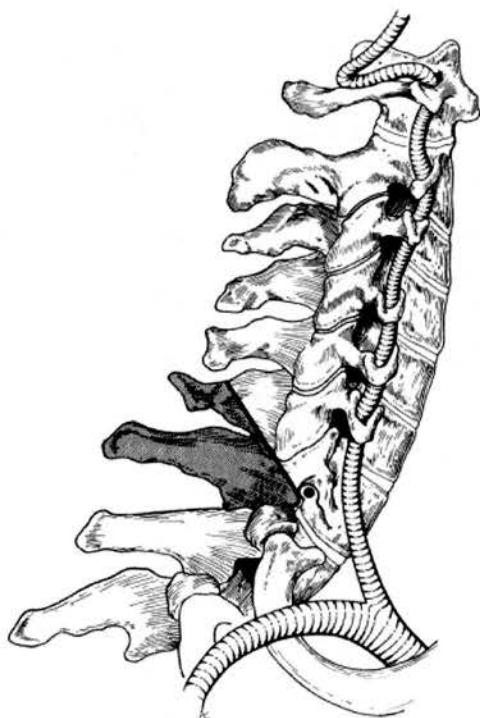


Fig. 19.14a

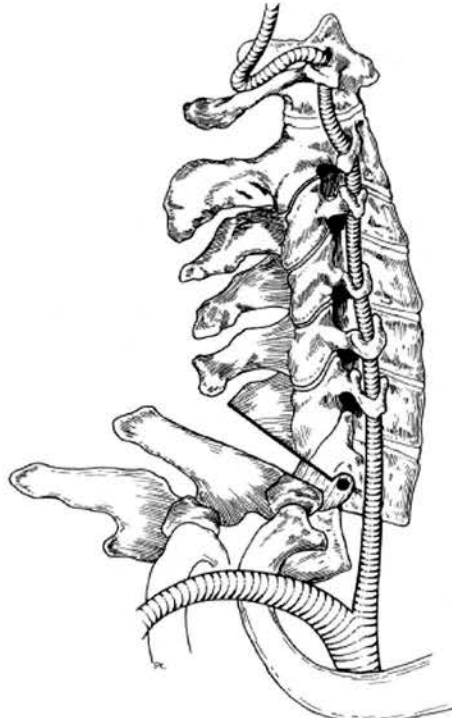


Fig. 19.14b

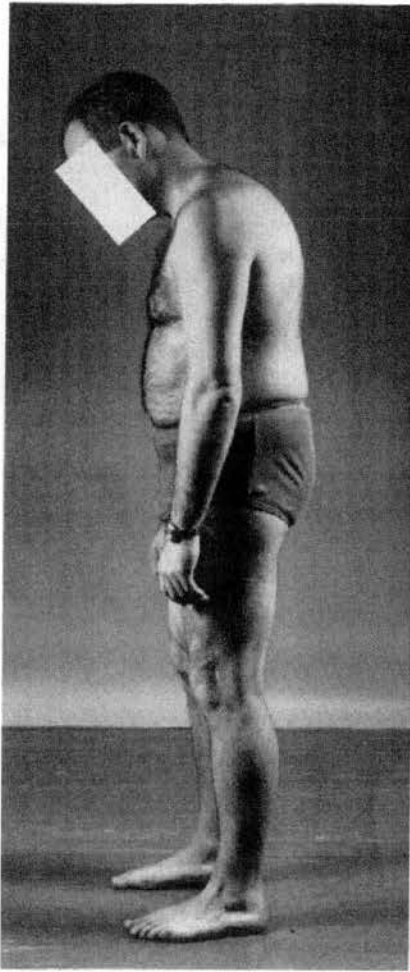


Fig. 19.15

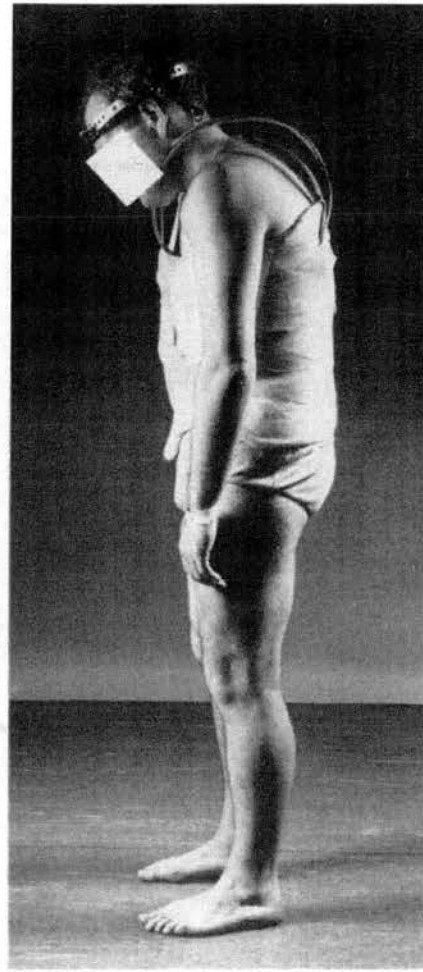


Fig. 19.16

A man with a severe flexion deformity at the cervicothoracic junction. Correction has been achieved by a cervical spine extension osteotomy performed at the level between the seventh cervical vertebra and the first thoracic vertebra. The ossified anterior longitudinal ligament has fractured and there is an opening of the anterior disc space. One year later the osteotomy has fused and the patient can see straight ahead.

To close the osteotomy the halo is firmly gripped and an assistant working beneath the drapes unlocks the hinges at the bases of the uprights supporting the halo and also releases the pistons within the supports. The surgeon who is gripping the halo then slowly and firmly extends the head and upper cervical spine while carefully observing the exposed dura. Correction usually occurs due to a fracture of the ossified anterior longitudinal ligament which may occur with a snap and an opening of the anterior disc space (Figs 19.17, 19.18). It is not necessary to carry out a preliminary anterior release of the cervical spine by cutting through the ossified anterior longitudinal ligament because this fractures very easily and it would be very difficult to approach this region anteriorly because of the severely flexed position of the neck with the chin resting on the chest. If the osteotomy has been cut correctly the initial resistance is easily overcome and the exposed dura wrinkles confirming that the spinal cord is being relaxed and not stretched. If the dura does not wrinkle, the lateral parts of the osteotomy should be inspected to make sure that there is no premature impingement of the bony structures posterior to the nerve roots. Once correction has been obtained the lateral masses at either side should come together without pinching the underlying nerve roots (Figs 19.13c, 19.14b). Once the head has been extended sufficiently, the hinges at the bases of the uprights and the pistons are locked in the corrected position.

The bone which was removed in the process of performing the laminectomy and osteotomy is cut into chips and placed bilaterally across the osteotomy site. Bone is not placed in the midline where it would impinge on the exposed dura.

Once the osteotomy is closed a wake-up test is performed and the neurology

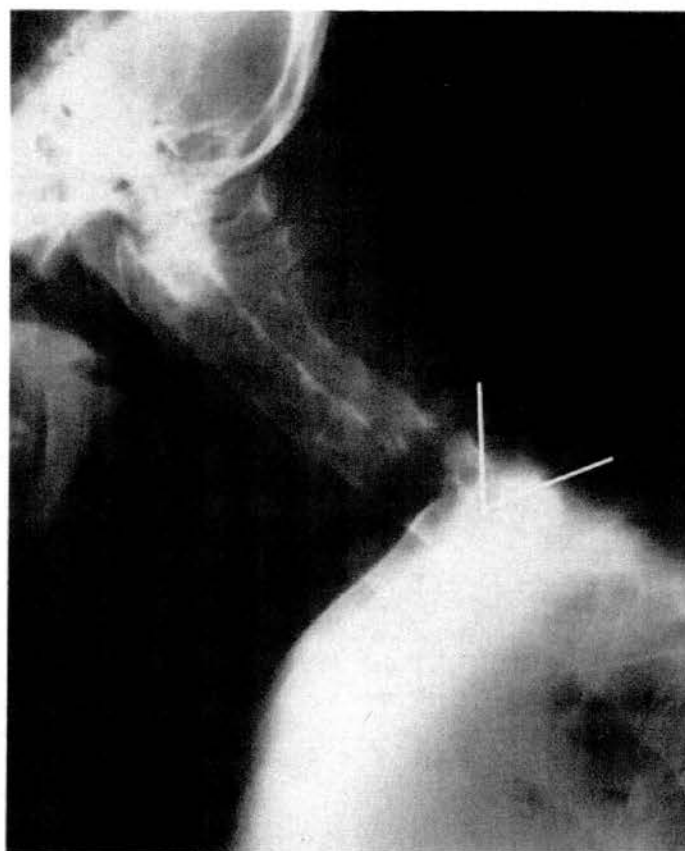


Fig. 19.17

in the lower limbs is checked. If there is a neurological deficit the hinges on the halo are released and the neck flexed forwards until the deficit recovers.

Postoperative care

After the operation the patient is nursed in a normal bed and turned from side to side. On the second postoperative day the patient is usually able to sit upright and take a few steps. It is not unusual to get intermittent radicular pain in the distribution of the eighth cervical nerve

roots in one or both arms. This pain usually occurs when the patient moves because although the neck is securely held there is still slight movement at the osteotomy site and this can cause irritation of the nerve roots. These symptoms usually subside within a few days.



Fig. 19.18

Once the patient is mobile and able to stand erect, the position of the head and his ability to look straight ahead are assessed. If the head is not in the correct position, it is possible to release the hinges on the uprights and make slight adjustments providing the patient is given adequate analgesia. In order to completely secure the halo and prevent accidental movement of the hinges, I usually apply two further supports to the halo on either side posteriorly and these prevent any accidental movement.

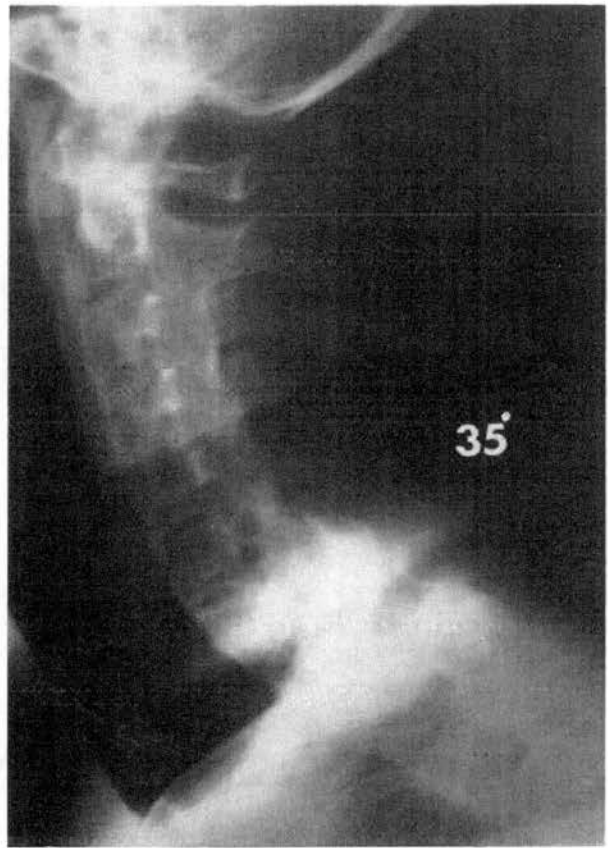


Fig. 19.19

The patient is usually able to leave hospital after 2–3 weeks and remains in the halo jacket for 3 months. At this time, X-rays are taken but these can often be difficult to assess with regard to the healing of the osteotomy. The halo jacket is usually removed at this time and the patient placed in a firm cervical collar which is worn for a further 3 months. After this the patient is left free (Figs 19.16, 19.19).

CONCLUSION

Spinal osteotomy either in the cervical or lumbar region of a patient with ankylosing spondylitis can be a difficult and potentially dangerous operation with a high incidence of serious complications. However, the majority of problems can be overcome by a meticulous surgical technique and careful attention to detail. A successful operation provides not only major functional advantages but also psychological benefits for the patient.

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